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# The Australian and New Zealand JOURNAL OF SURGERY

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## THE HERBERT MORAN MEMORIAL ORATION

### JOHN HUNTER'S TRADITION

By SIR GORDON BELL

*Dunedin, New Zealand*

THIS is the third lecture given under the aegis of the Royal Australasian College of Surgeons to commemorate the life and work of Herbert Moran, a graduate of the University of Sydney, a surgeon, scholar, linguist, writer and patriot. I met him first in 1909 when after captaining a successful tour of the "Wallabies" in Great Britain, he visited Edinburgh to take the Fellowship, and at that time he already bore the stamp of future distinction. On his return to Sydney to follow a surgical career he gave much attention to malignant disease at a time when treatment by radium was in its infancy but coming into prominence under the influence of the French and Swedish schools. He pioneered this method of treatment in Australia, won recognition as an authority in this field and made many trips to the Northern Hemisphere in a constant endeavour to add to his knowledge and efficiency in the management of cancer.

We in New Zealand have reason to remember him for he visited this country in the early days of the Cancer Campaign to give us the benefit of his experience and we recall his attractive personality, his professional enthusiasm, his wit, and his gaiety of spirit.

By one of those quirks of fate, an irony not uncommon in the case of doctors, he was destined to die in 1945 by the disease to which he had given greatest attention, and that in one of its most intractable forms.

Herbert Moran was deeply read in English and European literature and in the history of Medicine; and in his lifetime made a gift to the University of Sydney to found a prize for

an essay on "The History of Medicine and Science." It is therefore appropriate that the terms of this memorial address should postulate reference to the history of Medicine, and I propose to fulfil this condition by tracing the origin and growth of research in so far as surgery is concerned.

Time does not allow reference to those tremendous auxiliaries which have so greatly assisted the forward march of surgery and have themselves grown in remarkable fashion by researches contributed by a multitude of workers in the various fields. I refer, of course, to anaesthesia, bacteriology, radiology and later, biochemistry, blood transfusion and the methods of visualizing disease. Chemotherapy and the antibiotics require no mention for they are familiar to all, lay and professional alike.

Some would deny or denigrate the value of medical history but Winston Churchill has remarked, "The more we can look back, the further we can look forward"; and Charles Dickens makes Aunt Betsy Trotwood say to David Copperfield, "It's in vain to recall the past unless it works some influence upon the present." With these justifications for looking back and recalling the past we may now sketch the real birth of scientific surgical research two centuries ago and follow its progress.

### JOHN HUNTER AND THE 18TH CENTURY

In the year 1748 a redheaded young Scotsman, twenty years of age, John Hunter by name, set off from the family farm, Long

Calderwood, near Glasgow, to ride to London to join his brother. William Hunter, then thirty years of age, had already succeeded in establishing himself as an anatomist and man mid-wife and was a man of mark in London. He was therefore in a position to display that well-known and often derided Scottish trait of exercising a benevolent patronage and promoting the family interests. So well did he do it that it has been aptly said that without William Hunter there would have been no John for he set the pace in industry, opened the door to the treasures of nature and applied the spark to a kindred mind.

There is something symbolic in the picture of this raw youth riding off into the blue to challenge Fortune: like the immortal Spaniard though unattended by a Sancho Panza, John was to find many windmills at which to tilt, and many placid duckponds inviting stones. His journey from the farm to brother William's house in Hatton Garden occupied a fortnight, and his incredible labours over the next forty-five years set him securely among the Immortals. He died in 1793 on the same day that Marie Antoinette was beheaded and was buried in the vaults of nearby St. Martin-in-the-Fields. In 1859 his coffin was identified after a search lasting sixteen days and re-interred in the north aisle of Westminster Abbey. The long ride had ended.

With each successive generation, great human figures and their achievements recede and fade; memory is short and the new generation accepts things as they are as the natural order and all in the day's work. So the great man and his work come to assume an almost mythical character, commanding mostly lip service. Thus it is with John Hunter; with Lister who died only in 1912; and so it will be with our own Rutherford in his terrifying realm of nuclear-physics. But, perhaps some can echo with Herbert Moran, "I have been faithful to thee, Cynara, in my fashion."

For most it is both necessary and fitting to hold a recurrent interrogatory stocktaking and to ask: What is the Hunterian Tradition? Why are various aspects of his work taken as texts for the Annual Hunterian Orations? Why are the remnants of his museum left after the German onslaught on Lincoln's Inn

Fields in the Battle of London in 1941 rated among the chief treasures of the Royal College of Surgeons of England? The answer is short: John Hunter found surgery little more than a trade and forty-five years later left it a science, based on observation, experimental investigation and the inductive approach through the fields of human, animal and plant anatomy, physiology and pathology. His vision embraced nature and the range of his interests is illustrated in the contents of his museum, bought by the Government and transferred in 1800 to the keeping of the newly born Royal College of Surgeons of England. This contained 13,682 specimens: 7,795 in the field of physiology, 2,678 in pathology and 3,709 fossils and others. Furthermore, he preached a strange and revolutionary doctrine with its emphasis on cause being the thing that mattered in disease, not its end results: a sermon worthy of constant repetition two centuries later.

We may illustrate his approach by experiment to a problem and his genius for inductive reasoning by a brief reference to what is perhaps his most widely known contribution to practical surgery, i.e., the Hunterian ligation or proximal ligation for arterial aneurism. He had been interested in the growth and shedding of antlers and had tied the external carotid artery of a stag to observe the effect of cutting off the blood supply. The antler became cold but in a few days regained normal warmth and appearance, and on killing the stag, Hunter found that the smaller arteries above and below the site of ligation had enlarged and by their junctions restored circulation. In other words he had established the principle of collateral circulation with the inference that in man the smaller arteries are capable of rapid enlargement to take over the work of an occluded main artery, under the stimulus of necessity.

The next logical step was to apply this principle and the opportunity soon came in form of a patient with a large popliteal aneurism under his care in St. George's Hospital. Hunter tied the femoral artery above the aneurism in what is called Hunter's canal, with complete success. Proximal ligation, with the collateral vessels carrying on the circulation and allowing the aneurismal dilatation to solidify by clotting was quickly and universally acclaimed as a great advance.

This guiding principle of Hunter's mind is crystallized in his famous letter to a favourite pupil, Edward Jenner, the founder of vaccination. Jenner had written, posing some personal problem and Hunter replied, "Why do you ask me a question by way of solving it? I think your solution is just; but why think? Why not try the experiment?"

Hunter was a pioneer in tissue grafting and it is a pleasing fantasy to picture his delighted enthusiasm could he follow the recent chapters in the story of aneurism and the successful application of arterial grafts. The yeast of his tradition lives with a renewed vigour in this favourite field of his, and the wider applications of tissue grafting make a fascinating appeal to many present-day research workers.

John Hunter's habits of life and work and his colossal industry would have made even the traditional American hustler blench for his eighteen-hour working day was spent in unrelenting toil, dissecting, seeing private cases, performing his duties as surgeon at St. George's Hospital, teaching, adding to his museum, writing, doing his own thinking: his "filling of the unforgiving minute" would have rejoiced the heart of Rudyard Kipling. Even when he left the great house in Leicester Square for his country residence at Earls' Court (this rings strangely for modern London) he merely changed the scene of his labours for he there maintained a laboratory and a miniature zoo and aviary for his biological studies.

A note on Hunter's wife should interest us for man, however great, rarely travels alone. Anne Home illustrates the point that women have a keen eye for future distinction in a man, even if cloaked by a rough exterior. In her own right she was a woman of superior mind and education who wrote that haunting lament, "The Flowers of the Forest," the ballad, "My Mother Bids Me Bind My Hair," and the words of Haydn's "Creation," Joseph Haydn being an intimate friend and collaborator in the world of music. Her social activities accorded ill with John's devotion to work for she loved parties and was wont to fill her drawing room with assemblages of "musical professors, connoisseurs and other idlers."

Hunter himself was not without the national gift of pawky, grim humour, as witness his letter to his brother referring a patient for consultation (William already being a wealthy man). John wrote, "Dear Brother, the bearer is desirous of your opinion. I do not know his case. As he has no money and you don't want any, you will be well met." His delightful letter to young Jenner replying to a request to stand godfather reveals a quaintly playful affection. No wonder then that his pupils and a select group of friends loved him and called him "the dear man."

It has been said that a high priest's greatness may be measured by the stature of his acolytes. Hunter, supremely indifferent to pecuniary success, and valuing the "damned guinea" only as a laboratory aid, would doubtless have taken his pride and reward in the achievements of his pupils whose lives bridged the 18th and 19th centuries and whose work made English surgery of that period the admiration of the world. I refer particularly to Astley Cooper, Cline and Abernethy among the surgeons, to Edward Jenner, and, from across the Atlantic, William Shippen and Philip Syng Physick who carried back Hunter's teaching to become respectively the fathers of American anatomy and surgery.

It is safe to say that if John Hunter's researches had been confined to biology alone he would still have had his niche in the Temple of Fame, for his comparative studies and speculations foreshadowed by many years the work of Haeckel, Huxley and Darwin. Darwin's name has recently come into a happy and far-reaching relation with Hunter and his tradition. In 1928 Sir Buckston Brown, a Fellow of the Royal College of Surgeons, England, purchased Charles Darwin's old home at Downe in Kent and gifted it to the British Association as a national memorial. His generosity was then stimulated to the purchase of an adjoining thirteen acres on which he built and endowed the Buckston Brown Research Farm, in imitation of Hunter's country retreat at Earls' Court. This ideal place of research in country surroundings, generously endowed, is under the direct control of the Royal College of Surgeons, England. It will surely play its part in the forging of a "new Elizabethan era" in surgery under the aegis of that great institution.

So we may leave John Hunter to his eternal rest in the Abbey in company with those other immortals who have enriched the life of our country. He should live in the hearts of surgeons because he created the scientific approach and research and remains to this day, its greatest exponent and inspiration, at least in the English-speaking world.

#### THE 19TH CENTURY AND JOSEPH LISTER

John Hunter died in 1793 and though the next half-century saw a golden age in British surgery, decorated by a veritable galaxy of famous names such as Cooper, Abernethy, Cline, Brodie, Liston, Ferguson and Syme, it was not till 1865 that another star arose whose contribution was literally to revolutionize surgery. In just over a century since Hunter rode forth from Scotland to London, Joseph Lister journeyed by iron horse from London to Scotland to further his education in surgery under James Syme, the Edinburgh "Napoleon of Surgery." So was initiated that two-way traffic which promises to become more and more a feature of our modern post-graduate education.

There can be no greater contrast than between our two men of destiny: Hunter, the Scotsman, a country youth of meagre education, impetuous, overbearing, quarrelsome, ruthless in experiment and ruthless in opposition, with a cauldron-like mind, bubbling with ideas; Lister, the Englishman, a Londoner of gentle birth with the liberal education of easy circumstance, patient in opposition, calm and compassionate, dominated by one ideal, constant and relentless in its pursuit. Two things they had in common: first, a potent directing family influence; Hunter from an admired and brilliantly successful elder brother, Lister from inherited talent and scientific bent derived from his father; and second, each was moved by a compulsive spiritual force; Hunter by an insatiable scientific curiosity, Lister by the compassion aroused by seeing the suffering due to the impotent and frustrated surgery of his pupillage. One elevated surgery to a science, the other was to change the face of practical surgery so that we now speak of surgery before Lister and surgery after Lister: the Messiah had come, whose spark of genius was to save countless lives and to open the way to surgery as we know it to-day.

The story of Joseph Lister, the "contemplative Quaker," has been oft told but the fascination of his life and work still endures, for yet another "Life" appeared recently. My colleagues will bear with me if I recount briefly the pith of the matter. "Matter" is appropriate because it is the lay term for pus and it was the problem of inflammation in wounds and the attendant formation of pus that dominated Lister's mind. It was the almost invariable development of pus-forming inflammation and the subsequent death-dealing trinity of erysipelas, haemorrhage and gangrene that rendered accidental and operative wounds so dangerous and barred surgical progress (the day of the sulfa tablet and penicillin was far distant and bacteriology was still to come).

Lister's observations were predominantly clinical, or bedside, direct on patients; animal experiment came little into the picture if we except his testing of catgut as a surgical ligature and suture, and it would be fair to say that the hospital ward was his laboratory. None the less, Lister, the most humane of men, alive to the beneficent knowledge to be gained by animal experimental research, vigorously opposed an attempt to impose legal prohibition of such work. After many gropings for the source of inflammation and its exclusion by a variety of dressings, his prepared mind saw with inspired flash the significance of Louis Pasteur's work on fermentation and its application to the problem of wound infection. So was born the "germ" or microbic theory of inflammation, so followed the Antiseptic System of wound treatment, or Listerism, and so the way to the safe surgery of many hitherto perilous parts was laid open. The names of the great Frenchman, Louis Pasteur, and the Englishman, Joseph Lister, are linked in perpetuity across the Channel.

It in no way detracts from Lister's glory that the antiseptic system of surgery which prevented sepsis or wound infection, by a variety of chemicals like carbolic acid and mercurial solutions was before long replaced by our present day so-called Aseptic System. His system had solved the problem, the rest followed as logical refinements.

The scene of Lister's work see-sawed over some twenty years from Edinburgh to Glasgow and back to Edinburgh and he was professor in both. In lighter vein one may

comment that popularity in practice may be an unreliable guide to a surgeon's worth. Lister did not command popular success as a practising surgeon as witness his wife's playful or indignant reference to "Poor Joseph and his one patient." In 1877, at the age of fifty, he was invited to the Chair of Surgery at King's College, London. One can imagine the anxious discussions and heart searchings that went on between Joseph and his wife, Agnes Syme. Should they go? Should they leave the pleasant and distinguished position they held in Edinburgh, the scene of the inception of the great triumph, where they had met and most happily married? At last, characteristics of the man, and in spite of these considerations, and a petition from his students to remain, his sense of duty prevailed. With great reluctance he went south to preach his doctrine in London, and London received him with an even greater reluctance. The way of prophets is hard and their fate traditional.

Lord Lister died in 1912 and though Westminster Abbey was proposed, he was buried by his express direction beside his wife in West Hampstead Cemetery.

#### AFTER LISTER AND THE 20TH CENTURY

##### *The fruits*

A great scientific discovery and the opening up of a new country by an explorer are akin in their results. Then comes the host of eager, adventurous spirits who exploit, extend and enrich the primary discovery and so a new chapter is opened in world progress.

Let us see how this worked after Lister's conquest of sepsis, but first, let us define Research for that is the central theme of this address.

Research, so far as surgery is concerned, should have a broad definition. Liberally interpreted, it includes any useful contribution to knowledge, whether technical, whether accomplished at the bedside or the operating theatre, whether carried out in the laboratories of chemistry, physics, anatomy, physiology, pathology and bacteriology or in some special place labelled "Research Institute." Research has come in the minds of some people to have an almost mystical, remote and rarefied connotation and this interpretation is much to be deprecated.

Surgery is compounded of art and science, each open to advance by research in the wider sense. If we accept this view we can now survey some of the paths along which research and progress have travelled together since Lister, as the direct result of safe surgery. In pure science and in surgery one thing often leads to another and new vistas branch out from the parent stem, and new concepts are established. The vast expansion which went on in the last decades of the past century and in this, led to a similar expansion in types and field of research apart altogether from those tremendous ancillary aids mentioned earlier.

##### *Specialization, segregation and statistical research*

Various diseases came to operation in bigger and bigger numbers and, e.g., as early as 1884 Macewen of Glasgow could report 1,800 osteotomies with no serious complications. So it became possible to advance knowledge and improve treatment by statistical surveys or research by analysis. This may seem a purely mechanical application of book-keeping if no regard is given to the perplexities, thought and search for betterment animating the work which allows of statistical analysis.

Segregation of patients with particular diseases into special hospitals came more and more into use, as in the famous Queen's Square Hospital for Nervous Diseases. This provided surgeons with a concentrated experience, and the advances consequent on great experience, intelligently interpreted, constitute, if not research, its twin brother.

Furthermore, the now confident exploration of the body cavities came to reveal disease in its various stages, not only the terminal one seen at post-mortem examination hitherto. In this way the natural history of many diseases was laid bare in the living body, and this stimulated the physician, Clifford Allbutt, to coin the phrase, "the pathology of the living," which was later popularized by the surgeon, Lord Moynihan. To Moynihan, abdominal surgery has weightier debts than a striking phrase; notably, his artistry in painting word pictures of disease, and his early insistence on a scrupulous and gentle surgical technique backed by a gifted pen and great oratorical powers to promulgate his gospel.



### *Radical surgery in cancer*

In the great field of Cancer Surgery the outlook was immeasurably widened and the possibility of cure correspondingly improved. Take cancer of the breast as an example. Prior to 1890 many experienced surgeons admitted that they could not claim a single cure, but in the next five years as the result of intensive study, a correct welding of anatomy and pathology, based on a synthesis of previous individual researches on this problem, Halsted of Baltimore was able to formulate a logical basis of attack which was rewarded by greatly improved results. The same advance took place in other cancers, and by making anatomy (John Hunter's first love) serve its function as the fundamental basis for planned surgery, operations were devised to secure a radical extirpation of the disease. This involved much research on the part of anatomists, pathologists and surgeons, the keeping of accurate records and patient statistical review of results. Much has been gained and at the present moment progressive surgeons are testing extensions of radical surgery in the hope of better results. Yet another school of thought is testing, in the case of mammary cancer, whether a simpler operation combined with powerful radiotherapy may not yield improved results. Only the passage of time and the collection of massed statistics can give the answer. "Trying the experiment" still goes on. All this is trite to many of my colleagues but we should record our debt, and our gratitude, for the privilege of having worked in an atmosphere of optimism, not the deadly and shattering gloom of past days.

### *Endocrine system*

The fascinating territory of the ductless glands like the thyroid threw up new challenges to the surgeon and the physiologist. Kocher of Berne, the father of thyroid surgery, conducted an experiment on the grand scale in his assault on toxic goitre. His pioneer operations and experimental investigations towards the close of the century set the course for an immense amount of research and to an increasingly successful surgical treatment of goitre. Not only that, it focussed attention on similar glands with equally rewarding results. Truly, one thing leads to another.

### *The brain and spinal cord*

The brain and spinal cord were soon to attract the attention of the choicer spirits. Macewen of Glasgow is accorded by many of his compeers pride of place in opening the new era of neurosurgery. In 1879 he removed a frontal meningioma, five years before Godlee's classical excision of a true brain tumour. But it is in the realm of infections that he attained greatest distinction and his book, *Pyogenic Infective Diseases of the Brain and Spinal Cord*, published in 1893, was universally hailed as a classic. Macewen was cast in the heroic mould, an individualist and a true disciple as befitted a professor, who lived, taught and investigated a few miles from Hunter's birthplace. His contributions, especially on bone growth and grafting were the fruits of clinical observation of the highest order and the experimental testing of his ideas.

Horsley in London is the next commanding figure and added a new chapter by his strenuous operative work and his indefatigable investigation of brain function on anthropoid apes. Here we have the physiologist turned surgeon for Horsley, prior to his appointment to Queen's Square in 1886 had carried out more than 100 explorations of the anthropoid brain in his experimental studies on localization of function. During the eight-year period, 1884-1891, Horsley in collaboration with leading experimental physiologists, published eight noteworthy contributions to this subject.

The scene at the turn of the century now moves across the Atlantic and a young New Englander, Harvey Cushing by name, briskly takes the stage. Another individualist and perfectionist, Cushing's tireless industry, his development of a new technique in brain surgery, his perfect records and his masterly analysis of his vast collection of brain tumours in relation to sites, structure, natural history and individual clinical features, added the keystone to neurosurgery. That Harvey Cushing was early deeply imbued with the Hunterian tradition is evidenced by his organizing a "Hunterian Laboratory" in Baltimore about 1900 after his return from a *wanderjahr* in Europe, and later a similar research laboratory in Boston, the scene of his great work.



This story of surgical thought and research might be extended in many directions but my purpose is served if I have conveyed some idea of its wealth of diversity and of the sources which have created it.

The criticism may be offered that this is no more than a cursory and incomplete review of the advance of surgery. So it is but the advance of surgery and research are indivisible and I have endeavoured to retain the human interest as portrayed by some of the famous figures through two centuries of surgical history.

Research is open to all and though in this imperfect narrative I have woven the threads around some of our great heroes, research in its simpler form is within the capacity of any intelligent, industrious man. Who knows how the goddess Luck may reward the humble seeker?

The weightier contributions are likely to come from the man who, like John Hunter, has some consuming fire of curiosity ("Grand curiosity" as it has been called), or is, at least, an "anxious enquirer" as Macewen styled himself. The flash of inspiration followed by speedy and spectacular success is rare: Banting's discovery of insulin achieved in a few months is a good example. But *per ardua ad astra* is the more common fate and the investigator may labour like Jacob and find no Leah, "the tender eyed," or no Rachel, "handsome and well favoured," to reward him.

Reduced to the practical, clinical observation, recording and statistical review can still play a valuable part, pedestrian though it may be. For the rare, brighter spirits of superior mind, those with ideas and the zeal to test them, those who like Hunter, do their own thinking, ready access to a laboratory is essential and animal experiments play a great part. For the University professor of surgery in the modern sense, there must be adequate laboratory facilities, ample assistance and a generous budget if he is to fulfil his University obligations. For those devoted men who labour in the highly complex field of cancer research, terms somewhat reminiscent of Clause 6 of the Atlantic Charter are applicable: freedom of thought and approach and freedom from want, through a generous salary and pension.

### *Cost of research and the State*

The cost of research is as variable as its fields. It may be cheap and able to be carried on with the facilities in any well-equipped scientific department; but full-scale assaults on baffling problems, like cancer, are inevitably costly. Pure research must go forward along with objective research for a discovery of immense practical consequence such as penicillin, may come indirectly, more or less as a by-product.

In all this the State has its direct responsibility, and its attitude to, and financial support of the various national aspects of research, constitute one of the yardsticks of its enlightenment. It will be money well invested and we should remember that Louis Pasteur's work on diseases of wine and beer, on silkworm disease and on anthrax enabled France in an incredibly short time to pay off the indemnity exacted by the Prussians after the war of 1870.

If I may refer to a topical subject as bearing on the wider issues of national research in this great pastoral country, it is at least interesting to recall that in 1791, the eve of the French Revolution, the first Veterinary College was established in London. John Hunter appears in the list of vice-presidents and is generally regarded as having been the driving force behind it, and certainly as taking a leading part in drawing up a curriculum which insisted on a three-year course in anatomy, physiology, materia medica and pathology.

The New Zealand Government contributes the sum of £55,000 per annum to medical research in all its forms. Though this may sound a nice round figure the ideas of those submitting claims for Governmental financial support rarely accord with those who control the national purse strings. Taken in relation to the national income in these prosperous times, the amount is inadequate and the inadequacy is underlined if thought is given to the immensely greater sums expended on drugs in an endeavour to deal with the problems of disease at the wrong end. The perspective is out of focus, though understandable, but if New Zealand is to maintain and advance the position she has won in medical research, State support calls for a generous revision. It is of interest to note that the vote for medical research in the Commonwealth of Australia appears to be

roughly proportionate to our own, suggesting that officials' minds must think alike.

The New Zealand Branch of the British Empire Cancer Campaign controlling funds derived from public subscription and private legacies, contributes nearly £3,000 a year to the cost of cancer research. The whole picture of cost is heavily coloured by the greatly increased charges for equipment and salaries.

I think it would be fair to say that given the sinews of war, we can confidently predict that more and more of our graduates imbued with the enquiring mind, will come forward to shoulder the wheel of research.

#### *Research in New Zealand*

At present full dress research bearing directly on surgical problems is going on in the fields of cancer and endocrinology, the latter having developed as a logical extension of the original Thyroid Research Unit: also, with less direct surgical bearings, research being carried on in tuberculosis under the Travis Bequest, and pharmacological studies bearing on the medical treatment of high blood pressure, are both likely to cut down the amount of surgery necessary in these two great diseases. John Hunter regarded operation as a confession of failure, and surgeons will follow him in welcoming the replacement of surgery by other and better ways of treatment.

#### *Cancer research*

This had its beginnings shortly after the inception of the New Zealand Branch of the British Empire Cancer Campaign when in 1930 Dr. A. M. Begg was appointed Cancer Research Fellow to the newly founded Research Laboratory in the Otago Medical School. He was concerned principally with investigations on the mechanisms of spontaneous regression and disappearance in certain experimental cancers and the role of fibrous tissue reaction.

Dr. Bielschowsky, the present Director of Cancer Research, has worked principally on the conditions leading to the development of cancer in the thyroid gland and on the endocrine factors which bear upon mammary and ovarian cancers and cancer of the seminal vesicles. A point of historical interest is recalled by the investigations now being carried on in so many places on the endocrine

relationships of certain cancers which is regarded as a promising field for research. Beatson of Glasgow, as far back as 1896, pointed the way by showing that bilateral oophorectomy exerted definite retarding influences on mammary cancer. From these surgical observations of more than half a century ago has sprung an immense and widespread activity on the part of researchers and a renewed interest in, and practical use of, oophorectomy and adrenalectomy, and the application of sex hormones in appropriate cases.

Auckland, too, is inaugurating a campaign of active cancer research.

Another field of cancer research now transferred to the Medical School, concerns the physical aspects of radium and X-rays and their effects upon cells. This opens a field for speculation concerning a possible therapeutic agent against the cancer cell, analogous to the antibiotics.

Since the inauguration of the New Zealand Branch, consultation clinics have operated in various centres and have over the years made a contribution to the clinical study of malignant disease and statistical review thereon.

Finally, in other departments of surgery, the segregation of patients and the establishment of national units as in neurosurgery and in cardi thoracic surgery, have inaugurated useful research in these fields where closer cultivation yields reward.

The major forms of medical research touched on above, and others not mentioned, are an evidence of the growing maturity of this country and of the realization that we must contribute actively.

#### CONCLUSION

Though I have wandered far from the man whose memory we recall I think Herbert Moran would have approved. He was a man far above the average in his interest in literature and in the history of Medicine, recognizing his debt to our past great men, and drawing inspiration from them. In his work, devoted as it was in great part, to the study and treatment of malignant disease, he faithfully followed the Hunterian and Listerian tradition, probing the secrets of a fell, mysterious disease, curing or relieving where he could, and moved always by a compassionate heart.

## URETHRAL OBSTRUCTION IN CHILDHOOD

### THE USE OF URETHROGRAPHY IN DIAGNOSIS

By F. DOUGLAS STEPHENS

*Royal Children's Hospital, Melbourne*

WHEN symptoms of dysuria or enuresis suggest urethral obstruction in the infant or child, especially if the bladder is easily palpable, the elucidation of the type of obstruction requires special methods of investigation. Until recently, the sorting out of the level and nature of the obstruction has been done by endoscopic methods. The posterior urethra, a region of complicated embryology, fertile in its congenital anomalies, is an inaccessible structure in the male, requiring a range of miniature instruments for satisfactory inspection. Now it is realized, though perhaps not generally so, that great assistance can be obtained, both in infants and children, from the exact demonstration of the urethral contours by micturition cystourethrography. By this simple technique, exact diagnoses can frequently be made, usually without anaesthesia or instrumentation. In many cases, additional primary and secondary abnormalities in the urinary tract can be demonstrated to advantage, at the same time, and many suspected obstructions can be confidently disproved.

This paper discusses — (a) the technique and interpretation of this simple and accurate method of urethrography both in the normal infant and child and in cases of congenital urethral obstructions; (b) special studies undertaken to attempt to systematize the pathological anatomy and embryology of "valves" of the urethra; (c) the level of the obstruction in the urethra in Marion's disease of congenital origin, and the effect of spinal anaesthesia on the obstruction.

Part I deals with the technique and interpretation of the normal urethrograms, together with an anatomical study of the normal mucosal contours and muscular sphincters of the posterior urethra. Part

II describes individually the congenital obstructive anomalies of the urethra which were met with in this series — valves, strictures, Marion's disease, polyps, cysts of the bladder neck, diverticula of the urethra and meatal stenosis.

#### MATERIAL

In the series reviewed there were 175 patients whose ages ranged between newborn and puberty. These included 112 males and 63 females. On these, 241 micturition-cysto-urethrograms were made, some of which were post-operative. Of these, 103 showed normal urethrograms with or without upper urinary tract abnormalities. In 32, congenital urethral obstructions of various types were demonstrable. A third group of 40 urethrograms includes paralytic, or miscellaneous non-obstructive conditions of the bladder and urethra, none of which comes within the scope of this paper.

In addition, microscopic studies were made of the urethras of 3 boys and 4 girls to determine the extent of the segments of the urethra which were occupied by the internal and external sphincters and by the bulbocavernosus muscle. The urethra, pelvic viscera and bony pelvis were blocked in paraffin, not disturbed by dissection, and cut in serial sections of  $15\ \mu$  thickness. The musculature of the urethra was then plotted. These anatomical segments were then applied to their corresponding zones on the urethrograms.

The mural folds in the posterior urethra were macroscopically examined in 30 normal male specimens. This study was undertaken to ascertain the variations in arrangement of the inferior urethral crest, and to compare these structures with the folds which form "valve" deformities of the urethra.

### TECHNIQUES OF MICTURITION URETHROGRAPHY

The child or infant is placed in the supine position on the X-ray table. Non-toxic, non-irritating diadone compound diluted to 16 per cent. (Uriadone) is introduced into the bladder until full, or until the urge to micturate becomes intense. The catheter is then removed and cystograms are taken. These are followed by micturition cystourethrographic films. In children who are co-operative and who can start and stop micturition as required, antero-posterior, oblique and lateral views can usually be obtained. After completing these films, a final film is taken to demonstrate the presence or absence of residual content.

prove the presence of obstruction. Sometimes this can be obtained during the intravenous pyelogram series when the iodide solution in the bladder is sufficiently concentrated to demonstrate the urethra on voiding.

In girls, the true antero-posterior and oblique views are the most satisfactory. In boys, the true antero-posterior view causes superimposing of the wide column of iodide solution in the bulbous urethra on that of the membranous urethra obscuring the detail in both segments (Fig. 1). A near antero-posterior position may be obtained by packing a small folded hand towel under one buttock in order to tilt the pelvis and throw the bulbous urethra clear of the posterior

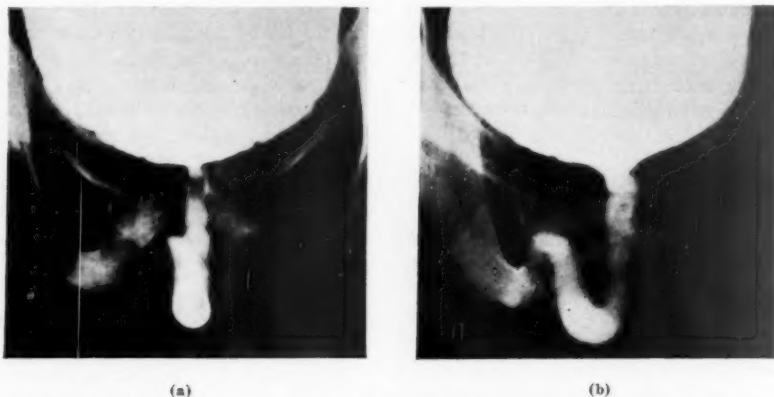


FIG. 1. R.H., aged 8½ years—normal urethrogram showing masking of membranous by the bulbous urethra in the true antero-posterior position in (a) and the appearance in the slight oblique (b).

Occasionally voiding may be possible only in the erect posture in which event satisfactory films may be obtained, although movement is difficult to control. Infants void spontaneously when the bladder is filled to capacity. Usually only one film can be obtained as micturition is so short in duration. If co-operation is not forthcoming, or the patient has chronic urinary retention, then it is unwise to introduce a catheter except at the time of operation. In this case resort may be made to manual expression under anaesthesia of the iodide solution from the bladder.

Quite often a single film of the urethra, taken during micturition, is enough to dis-

urethra. The particular advantage of this view in the male is that it shows well the membranous portion of the urethra, the verumontanum and the inferior crest and its terminal fins, whilst the oblique views show the bladder neck more clearly.

Absence of residual iodide immediately after micturition is a reliable guide to emptying efficiency, but the presence of residual solution, especially after intermittently arrested micturition, is not necessarily significant. In delayed films in cases with vesico-ureteral reflux, as discussed by the author in a previous paper (Stephens, 1954), residual urine, under these circumstances, is of little pathological significance.

## PART I

CORRELATION OF NORMAL URETHROGRAPHIC  
CRITERIA WITH NORMAL ANATOMICAL  
FEATURES

The occasional perplexing radiological findings in otherwise normal urethrograms necessitated a study of the various sphincter levels and zones in anatomical specimens of children. Close correlation of normal anatomy with the urethrograms has helped in explaining these features, and has offered some explanations in the problems posed by abnormalities in the urethra.



(a)



(b)



(c)

*Normal Urethrographic Studies. Male*  
(Fig. II)

Of the 103 subjects displaying normal urethrograms in this series, 61 showed normal and 42 abnormal upper urinary tracts.

The normal bladder showed a finely crinkled outline during micturition and the bladder outlet opened to a diameter proportionate to the force of effort of the bladder.

The posterior urethra, between the bladder outlet and the triangular ligament, showed a cylindrical column of iodide fluid, narrowing slightly anteriorly and posteriorly at the

FIG. II. P.T., male, aged 12 years—normal micturition cysto-urethrogram:

- (a) Slight oblique view showing to best advantage the urethral calibre and the detail in the membranous urethra.
- (b) Full right oblique view which shows the bladder neck and anterior urethra most clearly.
- (c) Lateral view which demonstrates the posterior margin of the bladder neck.

site of the ligament. The prostatic urethra formed a very slight bulge in the cylinder. At the midpoint of the posterior urethra, or slightly above it, the oval "filling defect" of the verumontanum on the posterior wall was seen. Below this the inferior urethral crest was sometimes visible as a central posterior longitudinal filling defect. Sometimes the terminal fins of the crest were seen diverging caudally to either side a short distance above the indentation caused by the triangular ligament.



The bulbous urethra, larger in calibre, turned anteriorly at right angles to the posterior urethra below the triangular ligament. The bulb extended to the site of attachment of the suspensory ligament and was approximately as long as the child's penile urethra.

The penile urethra extended from the suspensory ligament to the external meatus. Its diameter was narrower in the proximal portion. Inside the external meatus was the local dilatation of the *fossa navicularis*. The diameter of the normal external orifice distended during micturition almost to the calibre of the local dilatation just proximal to it.

The highest level of occlusion is the internal urethral meatus. The iodide column of the urethra is entirely eliminated from view, and the rounded even contour of the bladder base is clearly defined in the cystogram.

The second level lies at a region at or a little cranial to the verumontanum. The obliteration of the zone of the urethra below this level is demonstrable by instantaneous radiography at the moment of voluntary arrest of micturition (Fig. III). The upper limits of the iodide column in this zone become thinned in density and occlusion is complete caudally. This gradual occlusion in

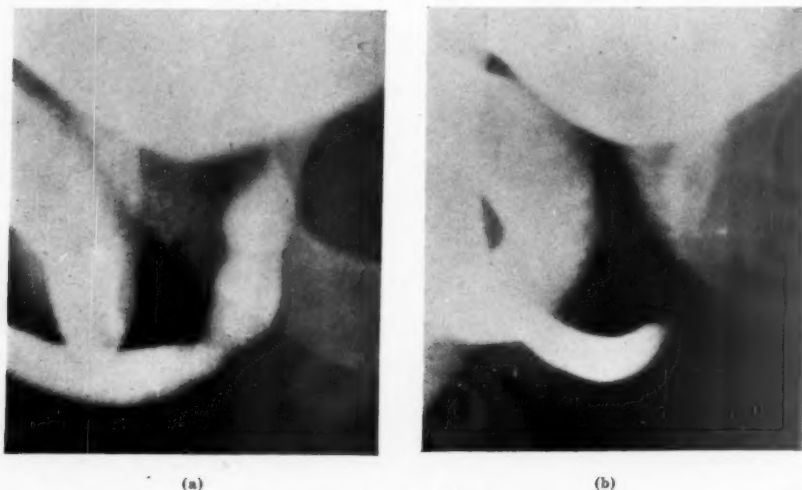


FIG. III. K.B., male, aged 11 years. Normal micturition cysto-urethrogram:

- (a) Posterior urethra and bulbous urethra during micturition.
- (b) At the earliest moment of arrest of stream by external voluntary sphincter contraction showing the upper prostatic urethra above and bulbous urethra below the contracted segment.

The column of iodide in the posterior urethra can be obliterated in two areas by voluntary effort. In routine micturition cysto-urethrographic series, only occasionally are films taken at the instant that the column is obliterated, with the result that the interpretation may be perplexing.

By correlation of the anatomical studies described below, and by reasoning deductively, these zones can be shown to correspond to the various sphincters of the urethra.

the male contrasts with the sudden complete elimination of the column in the female (Fig. VII). Presumably prostatic tissue in the posterior and lateral walls of the urethra in the male renders the sphincter mechanism less efficient at the upper limits. The lower limit of this zone is the upper surface of the perineal membrane, the level of which is sharply defined by the iodide still remaining in the bulbous urethra.

At the moment of occlusion of this zone by voluntary effort, the lower level of the



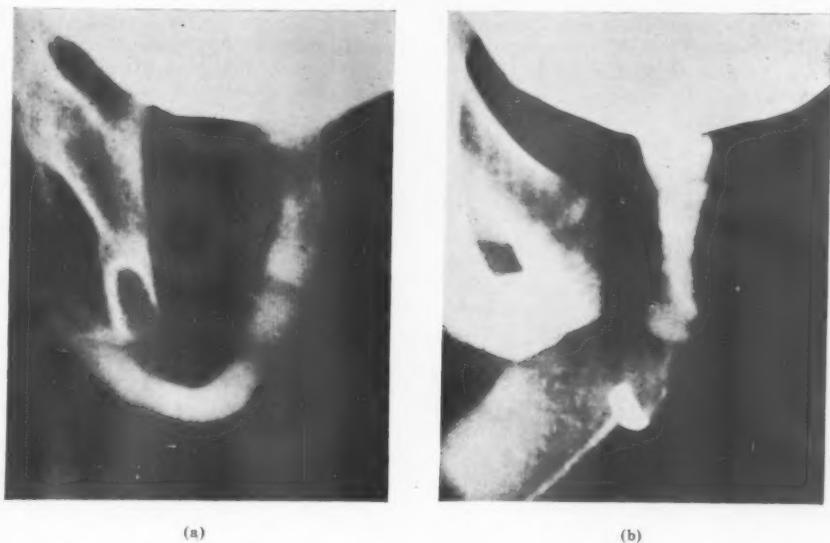


FIG. IV. B.M., aged 12 years:

(a) Normal micturition cysto-urethrogram.

(b) The lumen occluded by the action of bulbo-cavernosus muscle. Iodide is present in the collapsed urethra distal to bulbo-cavernosus.

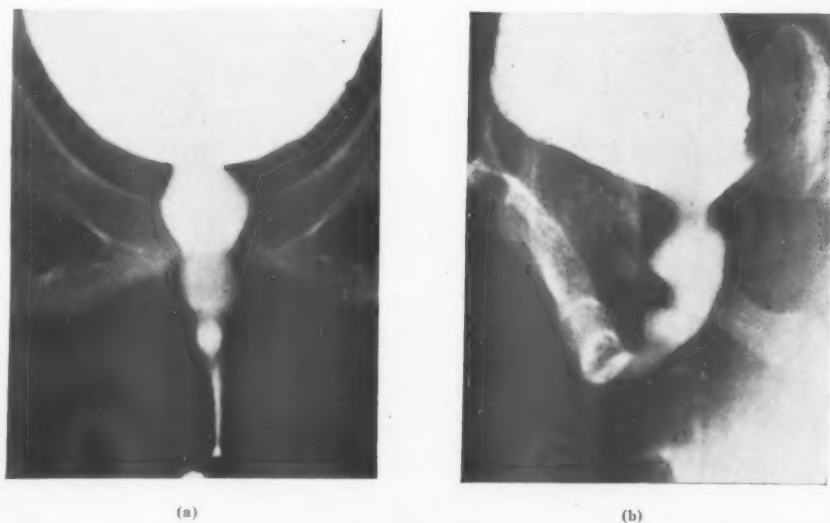


FIG. V. S.T., aged 7 years 2 months. Normal female micturition cysto-urethrogram:

(a) Antero-posterior view.

(b) Right oblique view. The indentation in (b) is presumably a peristaltic wave, more marked than usual.

upper zone becomes defined, since this zone remains momentarily visible as a cup-shaped extension from the bladder after cessation of micturition.

A third level of occlusion in the male lies at the site of the caudal surface of the perineal membrane. The zone of occlusion

extends distally from this point to approximately the region of the suspensory ligament. The column of iodide above retains its normal dimensions, but is cut off sharply at this level. Distally the iodide shadow in the collapsed lumen is momentarily narrow in calibre and sometimes fogged by movement of the penis (Fig. IV).

#### *Normal Urethrographic Studies, Female* (Fig. V)

The bladder in the female child showed the same crinkled outline during micturition as the male. The bladder neck opened to a diameter which varied with the strength of contraction of the vesical musculature.

The urethra during ordinary micturition shows up as an almost cylindrical column of iodide solution, sometimes with a gentle bulging of its upper and middle zones tapering to a narrower calibre quite rapidly at the external orifice. In many female children, the terminal orifice of the urethra is directed acutely anteriorly (Fig. V).

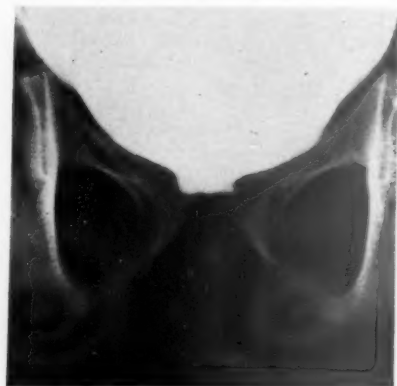
In some children whose micturition is excessively urgent, frequent and precipitant, the greater force of expulsion causes the bladder neck to open more widely, and the urethral lumen to distend to a greater calibre than normal (Fig. VI).



FIG. VI. P.E., aged 7 years 4 months. Enuresis. Micturition cysto-urethrogram showing widely open internal sphincter and dilated distal urethra.



(a)



(b)

FIG. VII. D.A., female, aged 6 years. Normal micturition cysto-urethrogram:

- (a) Showing upper bulge in the urethra demarcating internal involuntary sphincter zone.
- (b) The zone of urethral lumen obliterated by contraction of voluntary external sphincter and the internal sphincter still open.

In the female urethra, cessation of micturition by voluntary effort occludes only the lower much longer zone of the iodide column. At the instant of occlusion of this zone a short, sharply cut-off cup-shaped segment of the urethra amounting to one-quarter or one-fifth remains momentarily filled with iodide in free communication with the bladder. Finally this upper zone becomes invisible leaving only the smooth bladder outline.

In female children whose bladder contracted with great force, waves were sometimes seen on the anterior urethral outline suggestive of peristaltic activity (Fig. Vb).

#### *Anatomical Study of Posterior Urethra Micro-anatomy of sphincters*

Because of the great length of the iodide column in the urethral lumen which becomes obliterated by voluntary contraction of sphincters, a micro-anatomical study was made to identify and determine the extent of the external sphincter and the zone occupied by the internal sphincter.

In three male and four female post-mortem infant subjects the urethras were microscopically examined with regard to their normal relationships in the bony pelvis. Serial sections were cut in 15  $\mu$  thickness, and every tenth section was stained and examined.

The external voluntary sphincter in children was found to be more extensive than the short band in the lower urethra described in Gray's Anatomy (1954). The author's findings conform closely to those of Wood Jones (1902) who described and emphasized the long cylindrical conformation of the voluntary musculature of the urethra in the foetus of both sexes.

(a) *Internal involuntary sphincter.* The internal urethral meatus was surrounded by heaped up smooth muscle to form the internal sphincter. This occupied less than the upper one-quarter of the posterior urethra in the male, and of the whole urethra in the female (Fig. VIII). In the male it was entirely above the prostate.

It is particularly important to observe the extent of this sphincter on the micturition urethrograms, and to compare its appearance with those made from congenital Marion's disease (Fig. XIV).

(b) *External voluntary sphincter.* Little more than the lower three-quarters of the posterior urethra in the male and the whole urethra in the female was closely related to the external voluntary sphincter (Fig. VIII). The voluntary sphincter was a strong layer of striated muscle approximately equalling in thickness the two inner layers of smooth muscle. The muscle fibres were arranged obliquely, rising highest on the lateral and posterior aspects and intermingling with the lower and outer fibres of the internal sphincter. The fibres passed caudally and anteriorly across the midline to meet those from the opposite side. In the male, however, the circular fibres were thinnest posteriorly above the prostate; they gained direct attachment to the sides of the gland in the middle of the urethra, and formed a strong circular muscle in the lower urethra above the triangular ligament; in the female these circular fibres were more complete, extending to the level of the upper border of the ischiocavernosus muscle anteriorly and to a slightly higher level posteriorly.

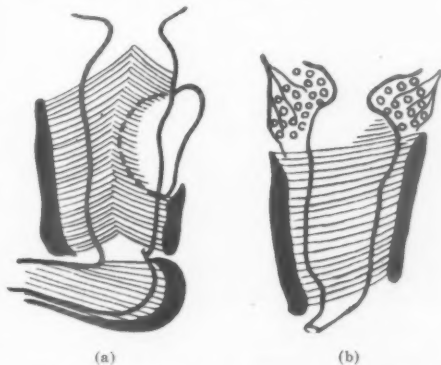


FIG. VIII. Voluntary musculature of urethra. (a)—Male. (b)—female. Note the proximity to the internal urethral meatus of the upper limit of voluntary muscle.

These urethrographic studies indicated clearly that the external voluntary sphincter was the sphincter which came into action first of all in voluntary arrest of micturition (Figs. III and VII). This observation is in accord with that of Denny Brown and Robertson (1933).

(c) *Smooth muscle of urethra.* Inside the voluntary muscle coat were two layers of smooth muscle, an inner longitudinal and an

outer circular. The outermost fibres of the circular layer were intimately related to the innermost voluntary fibres. The thickness of the involuntary muscle approximately equalled that of the voluntary coat. The function of these involuntary fibres is presumably peristaltic or detrusor, and furnished the explanation of the undulating outlines observed on cysto-urethrography (Figs. XII and VI). In both male and female, this musculature extended over the same segment of the urethra as the external voluntary sphincter.

(d) *Bulbocavernosus muscle in the male.* The fibres of this muscle encircled the posterior part of the bulb, arising posteriorly from the central point of the perineum, and more anteriorly diverging from the raphe along the caudal surface of the bulbous urethra to insert into the strong aponeurosis of the *corpus cavernosum urethrae*. Contraction of the bulbocavernosus muscle compresses the bulbous urethra and stops the flow of urine from the level of the perineal membrane. It can act as an accessory voluntary sphincter (Fig. IV).

#### *Macroscopic anatomy of the inferior crest of urethra*

Because valves of the urethra in children appear to be anomalies of the inferior part of the urethral crest, a special study of the normal crest was made.

The verumontanum and urethral crest were examined in thirty male cadaver specimens ranging in age from the newborn to nine years.

The verumontanum was situated on the posterior wall approximately at the midpoint or slightly above the middle of the posterior urethra. The inferior crest arose as a continuation of the lower end of the verumontanum, with or without a noticeable constriction at the point of origin.

In 18 cases the crest was a straight tapering ridge which terminated by dividing into 2 or 4 fins several millimetres above the spongy tissue of the bulb. These fins diverged laterally and distally in the membranous urethra to disappear at the level of the perineal membrane. In 5 cases, the appearance was similar to that described, but a fine central tapering ridge was continued

towards the bulb beyond the diverging fins. In 4 cases the crest terminated without dividing at the region of the perineal membrane. In two the central ridge was short and tapered rapidly to disappear on the posterior wall, but in these, two additional well marked right and left side fins, meeting at the point of origin of the central ridge on the verumontanum, were apparent. One further specimen showed only a doubtful left-sided terminal fin.

In some of the specimens examined, single or multiple very fine oblique mucosal ridges could be seen converging in a cranial direction on one or both sides of the central crest.

In all the normal cases, the central crest, as distinguished from valves, was present, even though in some it was shorter than usual.

## PART II

### URETHROGRAPHIC, EMBRYOLOGICAL AND PATHOLOGICAL STUDIES IN URETHRAL OBSTRUCTION

The particular radiological features of congenital urethral obstructions observed by the method of micturition cysto-urethrography are described. The term "valve" of the urethra embraces a group of anomalies which are individually studied, and which impel embryological explanation both of the normal crest and those anomalies involving variations in its development. The pathological anatomy of these anomalies, polyps of the urethra, diverticulae and meatal stenoses are correlated with the radiological findings. Doubts as to the exact level of the obstruction in the neuro-muscular inco-ordination of the bladder neck are implied by the radiographic observations and these are discussed.

#### *Valvular obstruction* (Corresponding to Type 1 of Young's Classification)

Young, Frons and Baldwin (1919) have adopted a classification to include all types of congenital urethral valves. These authors describe three types—Type 1, consisting of oblique postero-anterior mucosal folds below the verumontanum; Type 2, comprising postero-anterior folds diverging from the verumontanum towards the internal urethral

orifice; Type 3, appearing as a disc-shaped membrane or mucosal stricture with central orifice. They found that Type 1 was the commonest and Type 2 the rarest.

Type 1 is peculiar in that the mucosal folds develop a valve-like obstruction during micturition. The ballooning effect which occurs is similar to that which is seen in valves of veins. Type 3 obstruction is less valvular in effect, being caused instead solely by the smallness of the central orifice of the mucosal diaphragm or stricture.

and in 15 the fins merely met in the midline or joined the verumontanum directly (Fig. IXa).

The fins were firm unyielding structures which diverged from the midline posteriorly, traversing the lateral walls, to converge in the midline anteriorly at a more caudal level. Their tautness drew the mucosa of the lateral walls into two valve cusps whose selvedged edges lay contiguous in the sagittal plane. Their inflexibility contrasted with the expansile walls above, which dilated and hypertrophied.

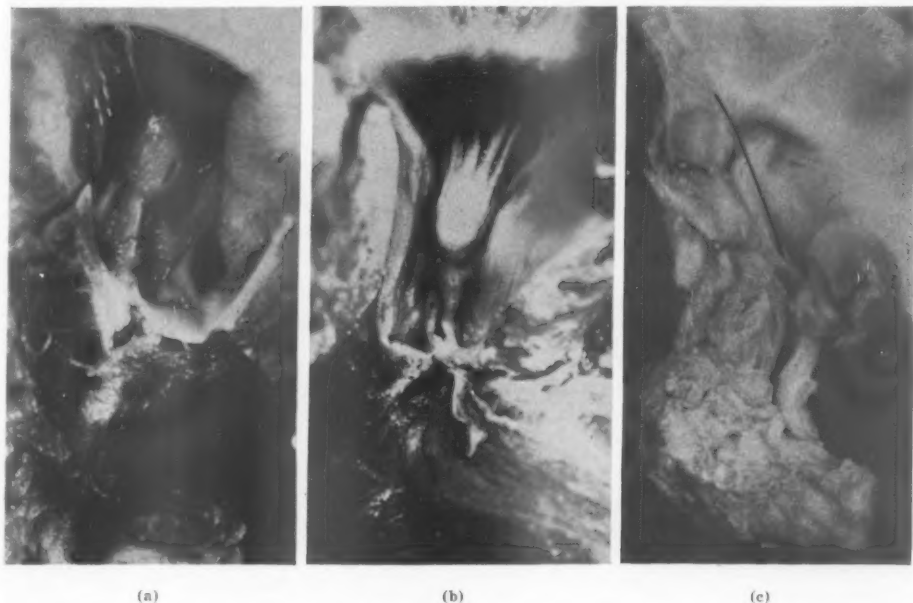


FIG. IX. Valves of urethra (viewed from the lateral aspect). These infant specimens show the sagittal slit-like orifice at different levels along the inferior urethral crest. (a) High, (b) middle, (c) low. In all the posterior urethra above the obstruction is widely dilated, and the verumontanum and crest are hypertrophied.

Congenital valves occur in males only. In my series there were 29 cases of this type of urethral obstruction. The valves were all located in that segment of the urethra lying between the verumontanum and the perineal membrane. All were intimately related to the diverging fins of the inferior urethral crest. Of the 19 post-mortem specimens which were investigated, the fins arose from the terminal end of a crest of approximately normal length in 3 cases (Fig. IXc), in 9 from a crest of medium length (Fig. IXb),

The slit-like urethral lumen between the valves was narrowed in side to side dimensions, and varied in antero-posterior diameter from approximately 0.3 cm. (Fig. IXa) to more than 1.0 cm. (Figs. IXb and X). The walls of the urethra caudal to the valves were not abnormally thickened.

The obstructive effect of the folds was in all cases valvular, but in some a true constriction of the lumen in both lateral and antero-posterior diameters was superimposed.

Secondary dilatation and hypertrophy of the urinary tract occurred above the valves. Some degree of renal damage had occurred from back pressure effects in most cases and in some hypoplasia of the kidneys was an accompanying feature.

The following differences between the conformation of the normal inferior urethral

(c) In valves the fins were thick unbroken structures which swept around and across the lumen to join together anterior to the urethral lumen. These contrasted with the brief delicate tapering ridges of the normal fins.

It would appear from our observations of these cases that valves in most cases

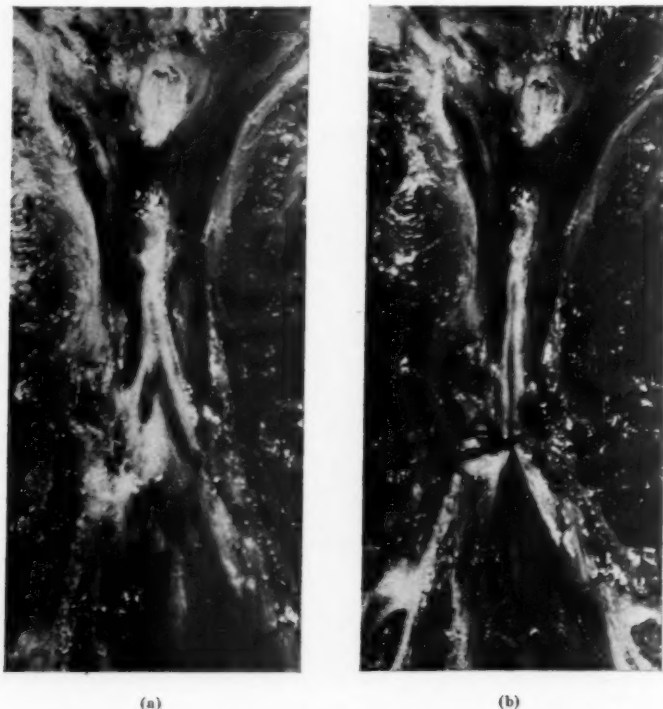


FIG. X. Valves of urethra (posterior urethra exposed by midline incision). M.S., aged 7 years. Specimen of posterior urethra showing (a) thick oblique fins of split crista diverging distally from the verumontanum around the walls of the urethra and (b) the anterior wall of the urethra reconstituted with a stitch demonstrating the long narrow lumen up which a catheter passes easily, but because of which in downward direction, urine is dammed back.

crest and its terminal fins and that which results in valves of the urethra were seen.

(a) The verumontanum, the crest and the fins were all larger and tougher than normal.

(b) In the great majority the inferior crest was abnormally short or absent. In only two out of 30 normals was the crest short, and in none was it absent.

represented more than hypertrophy of normal folds of the urethra. The fins were actually extended anteriorly to encircle the urethral lumen and the bifurcation of the fins was more commonly higher and at the expense of the inferior urethral crest.

This suggests that the normal crest was formed as a bilateral structure beginning as right and left contributions, and that the



terminal fins converged and fused in the midline to form the single central ridge.

The embryological significance of the normal folds and crest of the urethra is suggested by the appearance of the urethra of a male baby which I examined. He died at birth from congenital absence of the kidneys and all structures derived from the Wolffian and Müllerian ducts, namely the ureters, vasa deferentia, vesicles and ejaculatory ducts, and *utriculus masculinus*.

This specimen revealed a urethra of normal calibre in which the verumontanum was slightly more elongated than normal. The inferior urethral crest was deficient, and there were no fin-like folds radiating from any part of the midline of the urethra below the verumontanum. Prostatic ducts opened freely into the urethra. The *uvulae urethrae* and trigonal thickening were absent.

Confirmation of this observation by examination of other specimens of this rare abnormality is necessary. On this case alone, however, it can be postulated that the inferior crest, terminal fins and the occasional side ridges are vestiges of the terminal ends of the Wolffian ducts.

These ducts originally open into the cloaca close to the cloacal membrane (Keibel and Mall, 1912) at the 4.2 mm. stage. During the time of subdivision of the cloaca into anterior and posterior chambers, the terminal ends of these ducts are taken up into the urogenital cloaca so that the orifices of these ducts are transposed from their original location near the cloacal membrane to the site of the future verumontanum. From this point the ureters are then taken up into the bladder to the sites of the ureteric orifices.

The trigone is generally believed to represent the vestiges of these absorbed segments of ureter. By analogy the inferior crest and fins represent the vestiges of the terminal ends of the taken-up Wolffian ducts. The terminal fins demarcate the course of the right and left Wolffian ducts in the early migration from an anterior situation in the cloaca to a posterior position in the urogenital cloaca. In their cranial migration they lie side by side to form the urethral crest.

Valves of the urethra appear to be related to deficient integration of the Wolffian ducts

into the walls of the urethra, to abnormal locations of the original orifices of these ducts into the cloaca, and to an abnormal course of the terminal ends of these ducts.

No case of Young's Type 2 was encountered in my series. In many of my specimens of valves of Type 1, in which the prostatic urethra and bladder were hypertrophied, urethral folds were seen to diverge laterally and cranially from the verumontanum (Figs. IXa and IXb). These folds are probably similar to those described by Young *et alii* in their Type 2, but in my cases the folds were not obstructive.

In these studies, Type 1 is referred to as valves of the urethra, Type 2 is excluded for want of material, and Type 3 is referred to as urethral diaphragm or as mucosal stricture.

Tolmatchew (1870) believes that valves are simply hypertrophy of the normal folds of the urethra, and this is well-nigh true in the least common group in which the central crest is normal in length. In the majority of the cases studied in this paper, the folds, as enumerated above, are not only thickened but are abnormal in other respects.

Lowsley (1914) considers that the valve-like malformation which he describes is related to a defect in development of the Wolffian and Müllerian ducts. His explanation is based on a histological study of the normal and abnormal urethra. In the normal he found that these ducts entered the urethra at the verumontanum ensheathed in connective tissue which disappeared on the urethral floor below the entry of the ducts. In his specimen, this connective tissue persisted beyond the verumontanum gaining attachment to the walls of the urethra in such a fashion as to obstruct the lumen. His theory and my deductions are partly complementary.

#### *The technique and interpretation of micriturition cysto-urethrography in valvular obstruction*

Seventeen cases of valves of the urethra were examined by this form of radiography.

In cases of chronic urinary retention, it is unwise, because of the risks of infection, to catheterize the bladder except immediately prior to operation for relief of the ob-

struction. Better diagnostic films were obtained if the urethrography was performed in the conscious patient, though manual expression in the anaesthetized patient usually sufficed. As an alternative to catheterization in the anaesthetized patient, a smaller quantity of stronger iodide solution (50 or 70 per cent. Diadone) was introduced by direct injection above the pubis into the easily palpable bladder where the fluid became adequately diluted with the urine. Again, the urethrograms were obtained by manual expression.

The iodide solution, forced either by manual expression or by natural effort of micturition into the posterior urethra, sometimes dilated this structure to two or three times its normal diameter. With manual expression the internal meatus would open only slightly by passive distension, but with active micturition, it would open in proportion to the natural muscular effort. The dilated posterior urethra above the valves undermined the partly open internal sphincter, billowed out the anterior wall, and ballooned the lateral walls over the linear filling defects which represent the edges of the valvular orifice. Posteriorly the wall, reinforced by fibro-muscular and glandular tissue of the prostate gland, remained flattened except for the central enlarged verumontanum which appeared as a filling defect at approximately the middle, or a little to the cranial side. Occasionally reflux of the solution displayed the lumen of the ejaculatory ducts or utriculus masculinus.

The linear filling defects of the medial margins of the valves were visible, sometimes very faintly, in part or whole of their course. Their course was downwards and obliquely forwards, diverging from the filling defect of the verumontanum, or from the short posterior chest and converging near the anterior wall. The smooth, lower rounded contour of the dilated urethra was notched in one or two places by the filling defects of the valve margins (Fig. XII). The urethral lumen below the level of the valves was collapsed

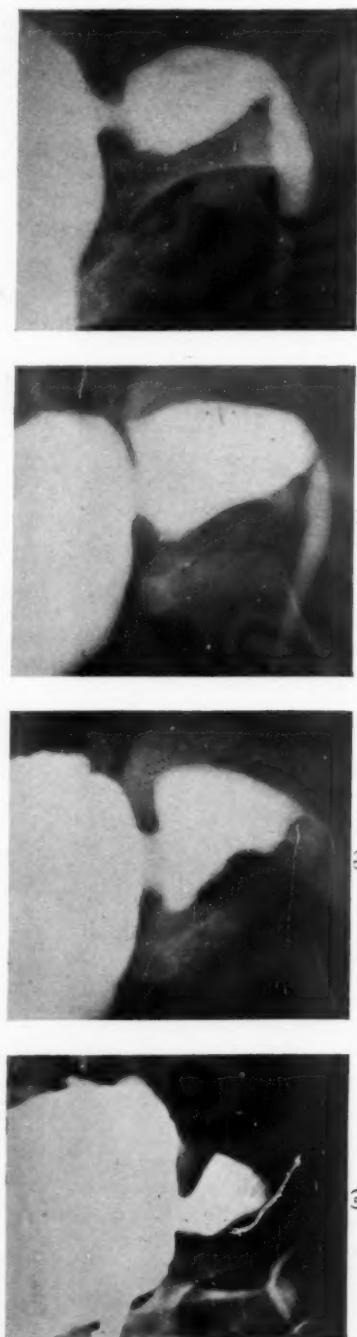


FIG. XI. J.M., 2½ years. Urethral Valves: (a)–(c) demonstrate gradual dilatation and filling of the bladder neck and posterior urethra to abnormally large dimensions during micturition. (d) Maximum dilatation and ballooning of the anterior wall and occlusion of the internal urethral meatus in region of junction of membranous urethra with bulb. (a) Moderate dilatation of prostatic urethra and filling defect of the enlarged verumontanum and narrow stream distal. (b) Rippling of anterior wall suggestive of peristalsis. (c) Maximum dilatation and ballooning of the anterior wall and occlusion of the internal urethral meatus in region of junction of membranous urethra with bulb. (d) Post-operative urethrogram at age of 4 years 4 months, 1 year 10 months after excision of valves by trans-urethral resection. Posterior urethra still dilated but no narrowing apparent. The internal urethral meatus is much reduced in diameter.

and showed as a narrow column of iodide: whether the whole or part of the membranous urethra was so delineated was determined by the level of the valves. The bulbous and penile urethral segments were diminished in calibre in proportion to the degree of obstruction above. The rounded lower margin of the enlarged iodide-distended urethra lying cranial to the fine streak of iodide representing the collapsed lumen of the urethra below, occasionally simulated the appearance of a mucosal stricture or of a diaphragm, but faint filling defects of the thick fins usually served to diagnose conclusively the valvular obstruction.

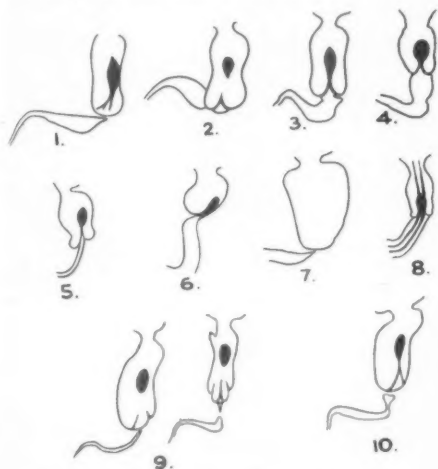


FIG. XII. Tracings of urethral valves in ten different urethrograms showing variations in X-ray patterns.

Upper urinary tract dilatation, sacculization of the bladder, tortuosity of the ureters and hydronephrosis of varying degrees were usually obvious, and vesico-ureteric reflux was seen in seven cases. In three of these an associated vesical diverticulum was also present with the ureter opening into it.

#### Mucosal stricture or diaphragm (Type 3 Young's Classification)

Post-mortem specimens were studied from 4 male infants. In these the obstruction was in the region of the junction of membranous urethra and bulb. In three the obstruction was a circumferential mucosal narrowing to a pin-point calibre (Fig. XIIIb). In the fourth specimen, the urethral lumen was partially

occluded by a diaphanous diaphragm perforated in two places (Fig. XIIIa). In all, secondary hypertrophy and dilatation was seen in the urinary tract above the obstruction and normality and collapse below.

The appearance of the verumontanum and inferior crest differed from that described in the valve deformities. In none of these three was there any evidence of hypertrophy of the verumontanum or deformity of the crest or fins. In the first case of mucosal stricture the verumontanum was represented by a depression in the posterior wall into which opened the ejaculatory ducts side by side. Emerging from the pit was a fine central crest which faded out just proximal to the site of the stricture. In the second and third cases the verumontanum was normal in size and the central crest was of cotton calibre terminating a little proximal to the obstruction. In all three the urethra was angulated acutely anteriorly at the level of entry of the ejaculatory ducts and utriculus masculinus (Fig. XIIIb).

In the case of the urethral diaphragm, the verumontanum and urethral crest were normal in size. The crest terminated immediately proximal to the diaphragm by dividing into two fine fins which formed the edges of the posteriorly situated small orifice. The second orifice was central and also very small in calibre (Fig. XIIIa).

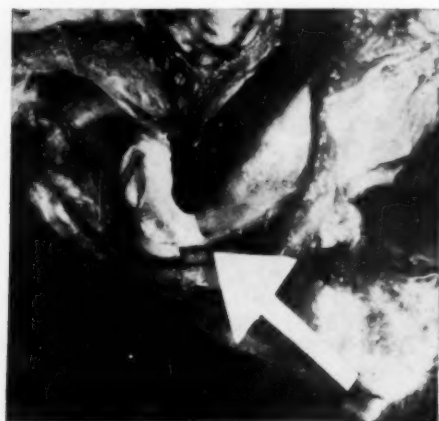
In three of these specimens, including that of the diaphragm, the ducts of Cowper's glands could be traced distal to the obstruction.

Strictures or stenoses of congenital origin occur commonly at the site of the bulbo-membranous junction (Lowsley and Kirwin, 1944). Bazy (1903) describes a clinical case of a valve-like obstruction in this region which he attributes to the persistence in part of the urogenital membrane. Young (1926) considers that Bazy's suggestion is probably the explanation of the mucosal strictures at this site.

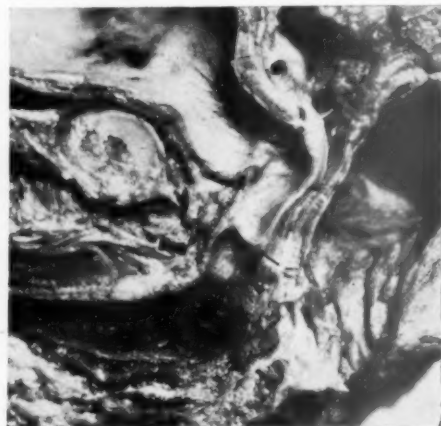
These four cases lend support to this embryological explanation, as all three are unassociated with abnormalities of the inferior crest, and doubts still exist as to the exact site of the urogenital membrane in the fully developed anatomy.

This theory, however, does not account for those even more rare strictures which have been described (Young *et alii*, 1919) in the supracollicular portion of the urethra. It may be postulated that these are fortuitous strictures common to all tube-like structures of the body, and bearing no special embryological significance.

cases of obstruction occurring at the region of the internal sphincter, yet showing no obvious anatomical or neurological cause. Marion (1933) describes the congenital and acquired types of this disease of the bladder neck. He considers that the congenital form, with which my subsequent remarks are concerned, is due to either muscular hypertrophy



(a)



(b)

FIG. XIII. (a) D.M., aged 5 weeks, urethral obstruction showing diaphanous membrane lying across the bulbous urethra immediately caudal to the junction with membranous urethra.

(b) Baby M., aged 3 months, urethral obstruction. Arrow shows level of mucosal stricture in the membranous urethra lying immediately above the junction with the bulb. Note also the acute angulation of the urethra at the level of the verumontanum.

In these three cases, micturition cysto-urethrography was not performed during life, but at the necropsies of the two children who provided specimens of mucosal stricture, the bladder and urethra were distended to capacity with iodide solution. X-rays demonstrated the narrowing caused by the stricture, the proximal dilatation, and the marked angulation of the urethra at the level of the entry of the ejaculatory ducts.

One child, who complained of some dysuria, was shown by cysto-urethrography to have a minor degree of narrowing at the junction of membranous with bulbous urethra. There was evidence of peristaltic undulation of the distended and slightly dilated posterior urethra. Urethral dilatation corrected the dysuria and disposed of the narrowing.

#### *Marion's disease of congenital origin*

The term neuromuscular inco-ordination of the bladder neck is used to denote those

similar to the congenital hypertrophy of the pylorus, or to absence or deficiency of the plexiform dilator fibres of the bladder neck.

In Marion's disease the bladder and upper urinary tract show all the typical secondary effects consequent on obstruction. The internal sphincter shares in the hypertrophy. The urethra is otherwise normal in appearance.

Unlike the valves and urethral membranes, the condition affected boys and girls, in our series in the proportion of 6:5. At the Royal Children's Hospital, Melbourne, over a period of four years, 2 cases of this disease were seen to 17 cases of valves of the urethra. These studies include a total of 11 cases—2 at the Royal Children's Hospital, Melbourne, and 9 at the Hospital for Sick Children, London. Seven were subjected to cysto-urethrography or cysto-urethrography. The findings in one boy, P.G., aged 2 years 9 months, are

recorded in detail. His investigations were of particular interest because of the special observations obtained by micturition cysto-urethrography.

### Case history

P.G., male aged 2 years 9 months. When 6 weeks old he developed pyelitis with painful micturition for which he received treatment. Recurrent tonsillitis followed during the age period of three to sixteen months. He always had a poor urinary stream which would stop, dribble, and start again during micturition. His appetite and weight-gain were poor. He was short and underweight (22 lb. 5 ozs. [9,214 grams]). His blood pressure was 160/110 mm. of mercury. His abdomen was full and the bladder was distended to the umbilicus and deflected slightly to the right by a solitary large left hydronephrotic pelvic kidney. The urine contained pus and proteus organisms. The blood urea was 105 mgms. per 100 millilitres. The micturition cysto-urethrogram revealed a trabeculated large bladder with urethral obstruction at or a little below the bladder neck, below which the stream was very fine (Fig. XIV). There was no reflux into the ureter, but subsequently through a suprapubic incision a No. 3 English gauge soft rubber catheter was passed freely up the left ureter into the left pelvic kidney. No kidney shadow was visible on routine intravenous pyelogram. At suprapubic cystostomy the internal meatus was found to lie deeply behind the pubis and the orifice was of apparently normal calibre though by digital palpation it was resilient. The orifice was dilated with sounds to 13/15. The urethra was dilated again two weeks and four weeks later but the suprapubic fistula would not close. At this stage micturition cysto-urethrography was repeated. Uriodone was introduced into the bladder through a fine catheter in the suprapubic fistula and the catheter was pinched off. Films were taken during active effort to micturate (Fig. XIVe). Iodide entered the upper one-quarter of the posterior urethra but would not pass further. With the reservoir held 18 inches above the bladder, no iodide would emerge from the external meatus. A low spinal anaesthetic was then administered. With the reservoir still held above the bladder a free urethral flow occurred as the spinal anaesthesia became effective. Fig. XIVf demonstrates the normal calibre lumen of the urethra when relaxed by the spinal anaesthesia. The child died from renal failure at the age of 3 years shortly after an operation on the bladder neck.

At post-mortem examination, the features already described were confirmed. In addition there was complete absence of both vasa deferentia and both vesicles. Prostatic tissue and ducts were present on the posterior urethra but the verumontanum occupied a more caudal situation than normal. A rounded mass of testicular tissue was present in the right half of the scrotum but the vas was lacking. Five lumbar and five sacral vertebrae were present and appeared to be normal. A large solitary kidney situated at the pelvic brim showed evidence of considerable hydronephrosis and pyelonephritis.

The cysto-urethrographic appearances of the bladder neck in this child are shown in Fig. XIV. The bladder neck lay closed when the bladder was

partly filled (Fig. XIVa). Micturition was attempted with the catheter still in the urethra and several films demonstrated the open bladder neck and an area of urethral contraction at a slightly lower level (Fig. XIVb). During the effort of voluntary micturition with the bladder filled to its maximum capacity, the region of the bladder neck became depressed in a cup-shaped manner and a fine stream of iodide emerged from its centre. Comparison of the levels of the resting bladder neck and of the "neck" at the time of voluntary micturition suggested that the internal sphincter opened widely and that the obstruction was met at a slightly lower level. A suggestion of the widely opened borders of the neck was gained from indentations in the iodide shadow at the sites where the cup-like depression evaginated from the bladder base (Figs. XIVb, c, d).

Further urethrograms were taken at a later date when the child was aged 3 years. Between the time of the first and second series of urethrograms a persistent suprapubic fistula had developed following the first suprapubic operation, when retrograde dilatation of the urethra was attempted. The fistula remained unaltered after subsequent urethral dilatations. All the urine emerged from the bladder through the fistula and with considerable force in the early stages when the track was healing in, but none came through the urethra. In the second series of cysto-urethrograms, the bladder was filled with iodide from a tube and funnel held approximately 18 inches above the level of the abdominal wall, through a catheter which fitted snugly into the fistulous track. X-rays were then taken during the effort of micturition before and after low spinal anaesthesia. No iodide was passed per urethram in efforts prior to spinal anaesthetic, but iodide did enter the upper end of the posterior urethra in approximately that zone surrounded by the internal sphincter. As soon as the effect of the spinal anaesthesia became apparent, the iodide passed through the urethra in a steady stream filling the urethra to normal dimensions to emerge from the external urethral meatus. This suggested that the obstruction lay in the musculature of the urethra which relaxed under the effect of spinal anaesthesia (Figs. XIVe and f).

The cupping of the bladder neck gave the appearance of extreme widening of the internal sphincter with incorporation of this zone into the lumen of the distended bladder rather than the general lowering of the bladder base which is ascribed to the relaxation of the levator ani musculature. The muscles of the urethra lying caudal to this sphincter are the external voluntary sphincter and the inner sleeve of involuntary muscle, either or both of which may be the cause of obstruction.

Cysto-urethrographic findings in six other cases of Marion's disease were of interest. In three of these children, the local cupping effect observed in the patient P.G., was even more apparent. In four cases, therefore, the appearances suggested that the internal



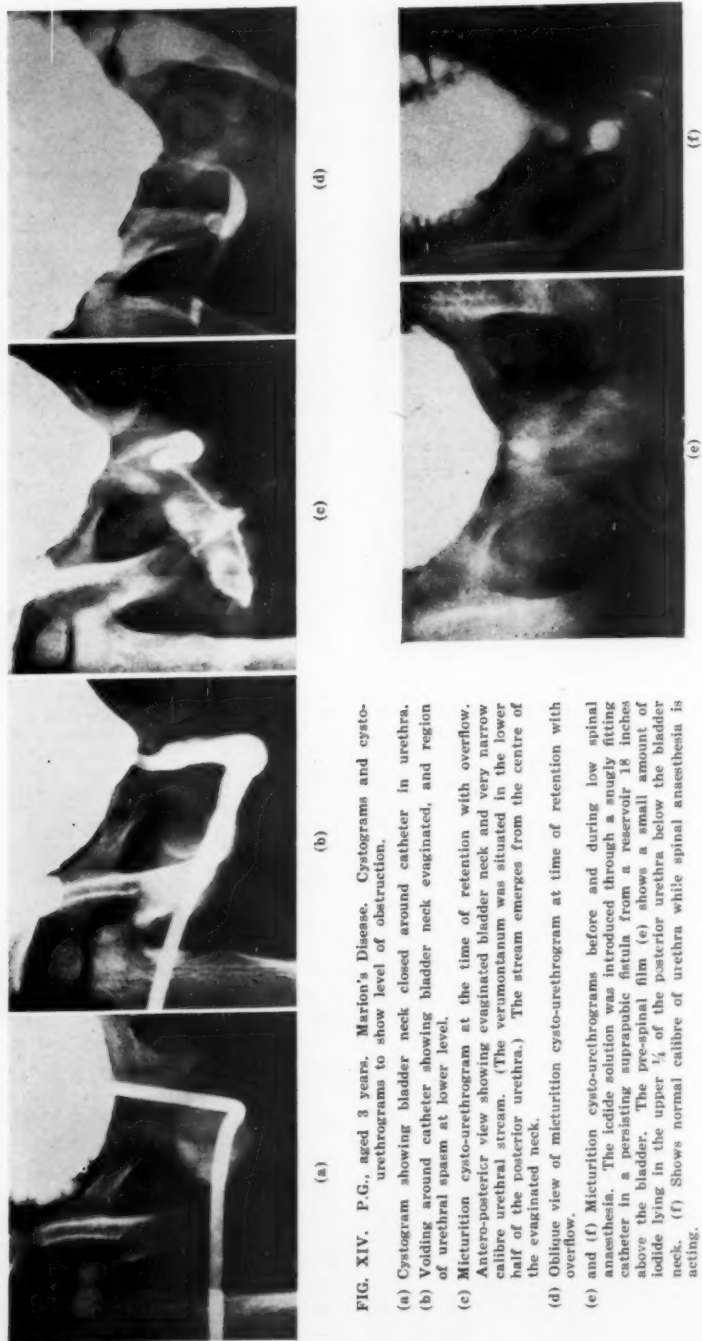


FIG. XIV. P.G., aged 3 years. Marion's Disease. Cystograms and cysto-urethrograms to show level of obstruction.

(a) Cystogram showing bladder neck closed around catheter in urethra. Antero-posterior view showing evaginated bladder neck and very narrow calibre urethral stream. (The verumontanum was situated in the lower half of the posterior urethra.) The stream emerges from the centre of the evaginated neck.

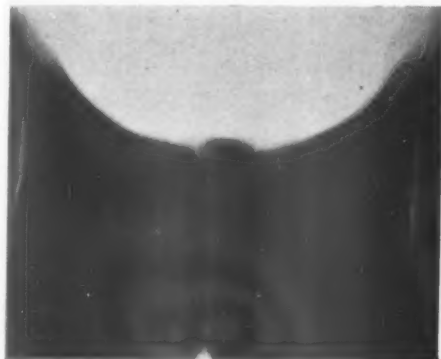
(b) Micturition cysto-urethrogram at the time of retention with overflow.

(c) Oblique view of micturition cysto-urethrogram at time of retention with overflow.

(d) Micturition cysto-urethrogram before and during low spinal anaesthesia. The iodine solution was introduced through a snugly fitting catheter in a persisting suprapubic fistula from a reservoir 18 inches above the bladder. The pre-spinal film (e) shows a small amount of iodine lying in the upper  $\frac{3}{4}$  of the posterior urethra below the bladder neck. (f) Shows normal calibre of urethra while spinal anaesthesia is acting.



sphincter instead of obstructing the urethra, widened to permit the passage of iodide solution, which was held up at a slightly lower level. Cystograms of two of these children are illustrated by Higgins *et alii* (1951). In the three other children cysto-urethrography showed no cupping effect.



(a)



(b)



(c)

FIG. XV. G.G., aged 5 years. Pedunculated urethral polyp.

- (a) Cystogram showing polyp in lumen of bladder at internal urethral orifice.
- (b) Micturition cysto-urethrogram showing oval polyp filling membranous urethra and filling defect of pedicle coursing cranially in midline.
- (c) Right oblique view of posterior urethra showing attachment of pedicle to verumontanum.

supposed; or is the obstruction at a slightly lower level? The relaxation under spinal anaesthesia observed in this one child (P.G.) indicates that the obstruction is neuromuscular in origin—not fibrous, prostatic or elastic; that the obstruction depends on outgoing impulses from the spinal cord; and that local or intramural reflex arcs alone were not responsible.

The muscles which come under suspicion at the lower level are the external sphincter or the inner sleeve of smooth muscle. Further studies, however, are necessary to confirm or disprove this hypothesis. Only when this level is more exactly defined can the hypertrophied internal sphincter be exonerated or incriminated.

#### *Polyp of verumontanum*

In two male children, aged 5½ years and 3½ years, a pedunculated polyp partially and intermittently obstructed the posterior

When, as on three occasions, I have inspected the bladder neck from within the bladder, I have noticed that it appears to lie in a deep depression in the base of the bladder; that the ureteric orifices are an unduly long distance from the internal meatus; and that digital palpation, and actual inspection of the orifice do not materially assist in determining whether the internal meatus is surrounded by the internal sphincter muscle or by the muscle of the urethra at a slightly lower level.

urethra. The polyps were approximately 1.0 c.m. in diameter attached to the region of the verumontanum by a stout stalk measuring approximately 1.5 cm. in length.

The polyp in the one case examined microscopically consisted of vascular connective tissue covered with transitional epithelium.

Cystography revealed a rounded "filling defect" of the polyp in the base of the bladder adjacent to the bladder neck. On

#### *Cysts of the bladder neck*

A large cyst caused obstruction at the bladder neck in one child. The symptoms of obstruction were insidious and overflow incontinence was noted at two years of age. At the age of four and a half years micturition cysto-urethrography was performed. This showed a large "filling defect" in the trigone region bulging out into the bladder, partially occluding the bladder outlet. A large bladder diverticulum was also apparent (Fig. XVI).



(a)



(b)

FIG. XVI. G.A., aged 4 years 8 months. Cysts of trigone obstructing urethral orifice and large vesical diverticulum, showing—

- (a) Filling defect of the cyst at the internal urethral meatus, and
- (b) Small diverticulum-like outpouching following marsupialization of the cyst, and absence of urethral obstruction.

micturition, the polyp on its stalk, fixed at the verumontanum, turned down and wedged itself in the lower membranous urethra (Fig. XV).

The posterior urethra and upper urinary tract were only very slightly dilated. The kidneys showed no evidence of damage due to back pressure. The urethra below the wedged polyp was small in calibre. Micturition cysto-urethrography subsequent to removal of the polyp and stalk revealed urethras of normal calibre and contour.

Apart from these cases of pedunculated urethral polyps arising from the verumontanum and probably of congenital origin, no case of enlarged verumontanum which obstructed the urinary flow was observed on cysto-urethrography.

Subsequent to operative marsupialization of the cyst into the bladder, micturition cysto-urethrograms were made. These displayed a free urethral outlet, an outpouching of the trigone at the site of marsupialization, and persistence of the vesical diverticulum. Pyuria persisted, and was finally cleared after removal of the diverticulum.

#### *Diverticulum of urethra*

Cases of intersex with enlargement of the utriculus or enlarged rudimentary vagina are excluded from this series. They did not cause urethral obstruction.

There were two cases of urethral diverticulum in this series. Both were situated in the penile urethra, and both caused some degree of obstruction.

In one child, aged five years, the diverticulum was small but a second malformation in the shape of valves of the urethra, was present also, and both contributed to the obstruction. The diverticulum became apparent on voiding. A tense rounded swelling, approximately 1.5 cm. in diameter, quickly interposed itself at the peno-scrotal junction under the perineal raphe, and slowly decompressed after cessation of micturition. The

than normal in calibre. Distal to the diverticulum the urethral lumen was collapsed and showed as a fine streak of iodide.

On dissection it was found that there was a fibrous stricture at the entry of the urethra into the diverticulum and at the exit there was a flap valve septum which became pressed against the roof of the urethra, as the diverticulum filled. This septum was not apparent on the post-mortem X-ray owing to the super-

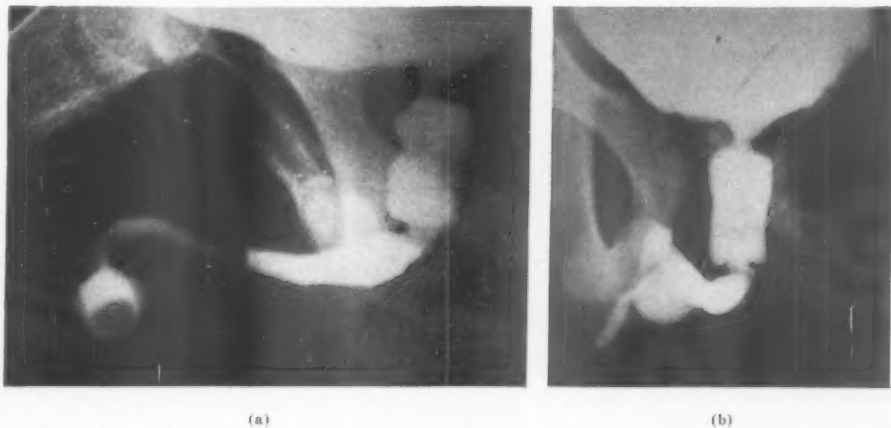


FIG. XVII. M.S., aged 5½ years. Valves of urethra and penile diverticulum. Valves in (a) show as faint forked lines diverging from filling defect of verumontanum and in (b) the notching of the lower outline of posterior urethra by the valve shadows. Diverticulum in penile urethra in (a) is pendulous sac and in (b) is fully distended encroaching on lumen of the urethra.

diverticulum, which was at first a pendulous sac, filled with iodide and encroached on the lumen of the urethra, adding to obstruction already produced by the valves (Fig. XVII).

The second patient was a newborn baby who died from urinary infection before cystourethrography was performed. The bladder was distended and palpable to the umbilicus. The diverticulum on each effort of micturition swelled into a sausage-like bag along the under side of the urethra in the anterior part of the scrotal sac. When milked in the direction of the external meatus, the sac could be emptied through the urethra.

After death the bladder and urethra were distended with iodide solution. The sausage-shaped diverticulum filled spontaneously. The urethra proximal to the diverticulum was normal in calibre for 0.5 cm. The bulbous and posterior urethra were slightly larger

imposed iodide shadow of the sausage-shaped diverticulum. Because of this masking of the septum, congenital fusiform dilatation of the urethra due to absence of the corpus cavernosum urethrae presents a cystourethrographic picture similar to that of the diverticulum. The urethral and upper urinary tract lack clinical and radiological evidence of obstruction where the dilatation is of the fusiform type only.

#### *Meatal stenosis*

This is the most common cause of urinary obstructions and is rarely demonstrated by urethrography as the diagnosis is obvious on clinical examination alone.

This condition occurs as a congenital or acquired lesion. The congenital form is due to the small size of the epithelial ingrowth in the glans penis and may be a local stricture at the outlet or a longer narrowing of the urethral lumen in the glans. It may be

associated with a hypospadias deformity when the patent meatus then assumes an abnormal position lying at the frenular region or more posteriorly.

The acquired type of meatal stricture occurs chiefly in circumcised babies. Napkin rash causes ulceration of the pouting mucosa of the urethra. This heals and fibrosis causes the narrowing.

On clinical examination during micturition the stream is abnormally fine and the penile urethra can be seen to become tense and expanded to abnormally large calibre.

On micturition cysto-urethrography the whole urethra is expanded by the iodide containing fluid to an extent which is proportional to the degree of muscular effort exerted by the bladder and the narrowness of the external urethral orifice. The stenotic meatus is apparent by contrast in the urethral calibre distal to the fossa navicularis, and by the narrow jet of iodide emerging as a stream. The posterior urethra in long standing lesions can distend to a calibre comparable with that in valvular obstructions. In the minor degree of narrowing, the urethrogram hardly departs from normal. Even six months after operative cure of the narrowing the distension is still present though not as large as preoperatively.

In those associated with hypospadias the whole calibre of the urethra is sometimes greatly enlarged out of all proportion to the degree of narrowing. In fact, even where no narrowing exists at all, the calibre of the whole urethra may be excessively large. Presumably the urethral dilatation is sometimes a developmental anomaly associated with the hypospadias deformity and independent of obstruction.

#### SUMMARY AND CONCLUSIONS

Over a period of five years, beginning at The Hospital for Sick Children, London, and continuing at the Royal Children's Hospital, Melbourne, a study has been made of 241 micturition cysto-urethrograms which were carried out on a total of 175 infants and children. In this study 32 cases of congenital urethral obstruction of various types were found. The particular advantage of this form of radiography is that, not only the obstructed, but also the normal urethral and vesical picture can be radiographically

visualized during the forceful effort associated with the act of micturition.

By the practice of this method, it was possible accurately to define the zones of the normal posterior urethra in relation to the internal and external sphincter, and to the bulbocavernosus muscle; and also to display the effective action of the external sphincter which is distributed over a greater length of urethra in children than is usually described for adults. This method also showed that, by comparison, the internal sphincter occupied a very short band. These zones were checked by corresponding micro-anatomical studies.

Correlation of the urethrographic criteria of valvular obstruction with the actual post-mortem specimens of this disease led to a more detailed study of the normal and abnormal urethral crests. Thus it was seen that valves were formed by the terminal thickened fins which diverged from the mid-line posteriorly and coursed obliquely to encircle the walls of the posterior urethra between the verumontanum and the perineal membrane. The higher the posterior origin of the valves, the shorter was the urethral crest, which in many cases was completely bifurcated when the valves arose directly from the verumontanum. Consequently, it could be postulated that valves were abnormalities of this crest and that the crest developed as a bilateral structure. Further evidence as to the origin of the terminal fins and inferior urethral crest was suggested by the study of one post-mortem case of a newborn male infant in whom the kidneys and all structures derived from the Wolffian ducts were lacking. In this urethral specimen, the verumontanum was slightly elongated, but the inferior crest and fins were absent. The absence of the fins and crest in this one case suggested that these structures may well be intimately associated with the development of the Wolffian ducts; that they represent the traces of the terminal ends of these ducts which during development are taken up into the urogenital sinus, transferring the orifices from their original sites near the cloacal membrane to the final position on the verumontanum; and that valves represent the terminal ends of these ducts which are deficiently integrated into the walls of the urethra, and whose course and terminal orifices are abnormally located in the cloacal walls.

Cysto-urethrography in cases of Marion's disease in children suggested that the obstruction to the flow of urine was at the level of the zone of the urethra which corresponded to the external sphincter and to that sleeve of involuntary muscle which lies internal to this structure. Urethrography further suggested that during voluntary attempts to micturate the zone of the urethra occupied by the internal sphincter might actually widen and seemed to become incorporated into the bladder as a cup-shaped appendage. In one special case, P.G., whose case history is recorded, cysto-urethrography demonstrated that the obstruction was immediately eliminated by low spinal anaesthesia; that the whole urethra dilated to normal dimensions; and that the obstruction was neuro-muscular, not mechanical.

Only one case of congenital mucosal stricture was diagnosed urethrographically but four other post-mortem infant specimens were studied. All five strictures occurred in the region of the junction of posterior urethra with anterior urethra. It is suggested that these mucosal strictures which occur most commonly at this site are remnants of the urogenital membrane.

Urethrograms of other congenital obstructions such as polyps, cysts of the bladder neck, penile diverticula and meatal stenosis are discussed.

#### ACKNOWLEDGEMENTS

This work was carried out over the last five years both at the Hospital for Sick Children, London, where I was privileged to study as Surgical Research Fellow for two years, and in the last three years at the Royal Children's Hospital, Melbourne. This personal study of these rare abnormalities of the urethra could not have been made but for the access to material so readily granted by members of the Medical Staffs of both these Hospitals. Some of the cases came under the clinical management of Mr. T. T. Higgins who permitted me to observe the children, assist with urethrography and other investigations, and in some to assist with the operations. In Melbourne I have had the help and guidance of Mr. J. G. Whitaker in the clinical management of cases. Many of the specimens examined were in the Pathology Departments of Dr. Martin Bodian at the

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#### REFERENCES

- BAZY, P. (1903), *Bull. Soc. Chirugiens (Paris)*, vol. 29, page 32.
- BRODNY, L. M. (1948), *J. Amer. Med. Ass.*, vol. 137, page 1511.
- DENNY-BROWN, D. and ROBERTSON, E. G. (1933), *Brain*, vol. 56, page 149.
- HIGGINS, T. T., WILLIAMS, D. I. and NASH, D. F. E. (1951), "The Urology of Childhood." London, Butterworth & Co. Ltd., page 121.
- JOHNSTON, T. B. and WHILLIS, J. (editors) (1954), "Gray's Anatomy, Descriptive and Applied." Thirty-first Edition, pages 597, 598. London, Longmans, Green and Co.
- JONES, F. Wood (1902), *J. Anat. (Lond.)*, vol. 36, *Proc. Anat. Soc.*, page 5.
- JORUP, S. and KJELLBERG, S. R. (1948), *Acta Radiol. (Stockh.)*, vol. 30, page 197.
- KEIBEL, F. and MALL, F. P. (1912), "Manual of Human Embryology." Philadelphia, J. B. Lippincott and Co., vol. 2, page 773.
- LOWSLEY, O. S. (1914), *Ann. Surg.*, vol. 60, page 733.
- and KIRWIN, T. J. (1944), "Clinical Urology." Second Edition. Baltimore, Williams & Wilkins Company, vol. 1, page 617.
- MARION, G. (1933), *Rep. Inter. Soc. Urol.*, vol. 1, page 392.
- STEPHENS, F. D. (1954), *Aust. N.Z.J. Surg.*, vol. 23, page 197.
- TOLMATSCHEW (1870), *Virchow's Arch. Path. Anat.*, vol. 49, page 348.
- YOUNG, H. H., FRONTZ, W. A. and BALDWIN, J. C., quoted by YOUNG, H. H. and DAVIS, D. M. (1926), "Young's Practice of Urology," vol. 2, page 86. Philadelphia, W. B. Saunders Company.



## MULTIPLE NEUROFIBROMATOSIS

By BARTON VENNER

*Aide*

**E**VEN before von Recklinghausen's classical description in 1882, the condition of multiple neurofibromatosis had attracted considerable attention. There has been a great host of contributions since and the literature of the subject has by now become indeed an *embarras de richesse*.

The hereditary nature of the disease was established by Alexis Thomson (1900) in his monograph. Preisser and Davenport (1918) showed that it is transmitted as a Mendelian dominant, so that it passes directly by either parent to the offspring, about half of whom can be expected to show the stigmata of the disease.

The presence of the diverse lesions was, of course, first recognized but Parkes Weber in 1909 clearly described some of the less obvious clinical manifestations—the *formes frustes* of the disease. More recently still, Holt and Wright (1948) have put on record the various skeletal lesions which can be detected radiologically.

### CLASSIFICATION

The term multiple neurofibromatosis is applied to a disease in which patients present with diffuse and nodular proliferations of the ensheathing tissue of nerves; these are either small or large and occur in association with a patchy pigmentation of the skin. Clinically, such cases are most often seen with multiple cutaneous or subcutaneous tumours, and the so-called *café au lait* patches of skin pigmentation.

The features may vary widely from the well-known type and the following general groups of neurofibroma may be recognized:

1. Multiple neurofibromatosis.
  - (a) Cutaneous (*molluscum fibrosum*).
  - (b) Generalized.
2. Plexiform neurofibroma.
3. Elephantiasis neuromatosa.

### 4. Solitary neurofibroma.

- (a) Cutaneous.
- (b) In other sites.

### AETIOLOGY

The condition must surely be a developmental one and the association with other congenital defects is frequent. In many cases a family history is clearly apparent, but sporadic cases are by no means uncommon. It is well established, too, that the first appearance of the lesions is often synchronous with puberty, pregnancy or the menopause, so as to suggest that there may be an endocrine influence (Herrmann, 1950). An increase in the number and size of the tumours at the times of successive pregnancies has also been observed (Sharpe and Young, 1937). See Case 2.

### PATHOLOGICAL FEATURES

The condition has been described as one of "neuro-ecto-dermal dysplasia" and has a similarity to tuberose sclerosis, in which multiple small cerebral cortical nodules are associated with cutaneous abnormalities of a vascular or pigmentary nature.

Neurofibromatosis may be regarded as a disease which involves the sheaths and enveloping membranes of the nervous system. It will be recalled that the peripheral nerve is composed of a number of myelinated or unmyelinated axons, each surrounded to its ultimate termination by the sheath of Schwann, a derivative of the neuroectodermal cells of the neural crest. Binding the bundles of nerve fibres together, and ensheathing the nerve trunk, is the connective tissue of the endoneurium and perineurium; as each nerve emerges from the dural sheath of the central nervous system, arachnoid ceases and the perineurium takes over its function.

Although the most striking feature of the disease is the nodular lesion of nerves, there is also a diffuse thickening of nerve trunks, both large and small, and the tumour-like

nodule is only a local exaggeration of the process (Thomson, 1900). The "neurofibroma" is a spindle-shaped swelling on an already thickened nerve trunk. In the skin, the fibromatous process may produce a considerable diffuse thickening, for the proliferation of the connective tissue of fine nerves in the dermis results in an increase in the thickness of the layer.

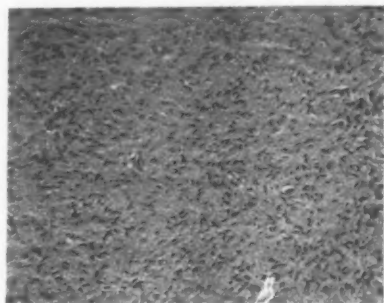


FIG. I. The loose arrangement of small spindle-shaped cells in a cutaneous neurofibroma. No palisading or whorled effect is present. (x 175)

Autonomic and somatic nerves may be alike affected; fibromatous nodules in skeletal muscle and the walls of hollow viscera have been noted (Beattie & Dickson, 1943).

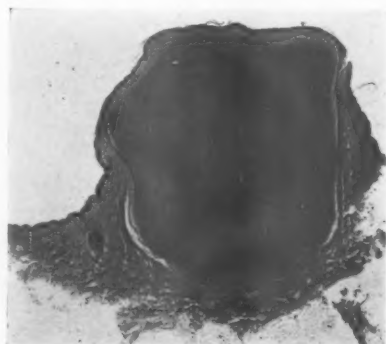


FIG. II. Neurofibroma in the dermal layer of the skin, producing a sessile tumour of the "molluscum" variety. (x 12)

In an advanced degree, the fibrosis may manifest itself as a plexiform neurofibroma or as elephantiasis neuromatosa. In the former, there is a localized aggregation of the thickened, nodular and interwoven nerves,

producing an irregular tumour in the subcutaneous tissues; in the latter, the presence of a large mass of neurofibroma tissue related to the cutaneous and subcutaneous nerves produces a coarse thickening and enlargement of a part. Both of these last-mentioned conditions are rare.

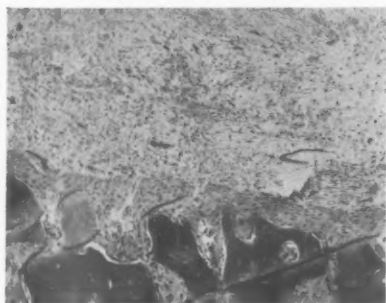


FIG. III. Section from the right radius of the patient shown in Fig. IV, at the site of a surface depression or "pond." (x 90)

The individual nerve sheath tumours have been variously called neurofibroma, neurilemoma, Schwannoma or perineural fibroblastoma. Many more variants have from time to time been suggested but this list includes those which most often find acceptance. Controversy exists as to whether these tumours arise from the ordinary connective tissue elements of the nerve, or are derived from the cells of neuroectodermal origin of the sheath of Schwann. The demonstration by Masson (1932) that all pigmented naevi are true "nerve-ending neuromas developing from the peripheral sensory apparatus" is of some significance in view of the common association of pigmentation in the disease, and histologically the pigmentation of von Recklinghausen's disease is identical with that in the pigmented naevus. More recent work, too, on the origin of pigment-forming cells and the migration of the "dendritic cells" from the neural crest, may have some bearing on the condition.

The histological appearance of the lesions is typical. There is a fibrosis, both localized and diffuse, in the nerve trunks, due to a great increase in the cells and fibres of the endoneural and perineural connective tissue. The origin from a particular nerve is not always apparent. The tissue has a tangled

reticular appearance, with interwoven strands of fibres and bands of spindle and elongated cells, sometimes dense and sometimes loosely packed, and often with abundant small vessels (Fig. 1). The elongated cells may be curled into a whorled appearance, as seen in meningiomata, or there may be a regimentation of the elongated nuclei side by side to give a palisade effect, seen best in the typical neurilemoma.

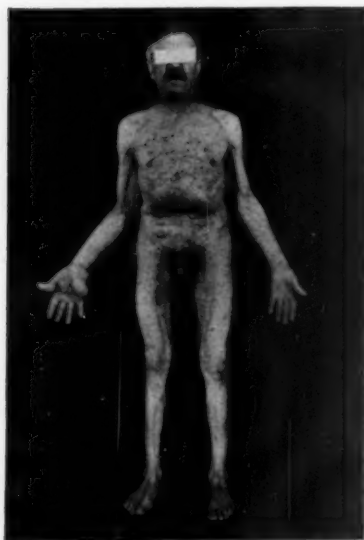


FIG. IV. Extensive cutaneous lesions of molluscum fibrosum. Note the large subcutaneous tumours deforming the right hand.

In the nodular lesions, the nerve fibres may be seen to be displaced to one side of the tumour or sometimes to pass through it without interruption. Apparent encapsulation is sometimes seen but more usually the nodule is continuous with a fibrosis extending along the nerve.

In the dermis, the fibromatous tissue forms discrete nodules (Fig. II), or a more diffuse infiltration. Bone involvement from contiguity of the neurofibromatous masses results in erosion of the cortex, localized cyst-like rarefactions and an elevation of the periosteum with new bone formation (Fig. III).

### CLINICAL FEATURES

It is likely that many of the lesser degrees of this condition are not recognized and, as has been the case in congenital bowing and pseudarthrosis of the tibia (see below), its association with some congenital defects may remain undetected.



FIG. V. The molluscum lesions are most numerous on the trunk.

The patient typically presents himself on account of superficial tumours, which may be few and only slightly disfiguring, or numerous and disabling. Over 4,000 tumours have been counted on one patient.

(a) The molluscum type is most conspicuous.

The trunk and limbs may almost be covered with soft, sessile or pedunculated tumours, from pinhead size up to two or three centimetres (Figs. IV and V). They present no precise regional distribution, although the trunk is often most heavily affected. The lesions are flesh-coloured, not pigmented, and the more vascular ones may be bluish and compressible, and resemble haemangiomas. They are sessile or pedunculated, and the overlying skin is closely attached to the tumour, which is, in fact, in the dermis. They are not normally sensitive to touch nor is there any sensory change

in the region. The clinical features of this form of the disease are illustrated by the case records that follow.

### Case 1

This patient, a married woman aged 33 years, was admitted for the treatment of an essential hypertension, with severe headache and nausea. The history elicited that following each successive pregnancy she had been getting "warts and lumps" all over her body.

Examination showed that the face and trunk had numerous pedunculated and sessile fibrous nodules in the skin. There were numerous large and small patches of *café au lait* pigmentation, but no deeper nodules were felt, and there were no other significant abnormalities.

This patient represents a common type of the condition often encountered, in varying degree of manifestation, in hospital patients as an incidental finding, and no doubt exists in proportion in the general population.



FIG. VI. *Café au lait* pigmentation, consisting of the typical larger patches, and a more diffuse speckling. The subject is one of affected twins; their manifestations are alike in general type only. The mild scoliosis can be seen.

### Case 2

N.S., a woman aged 49, is the daughter of the patient shown in Fig. IV and has twin daughters, one of whom is shown in Fig. VI. She complained of the grotesque appearance of several pedunculated nodules on her face, and it was soon elicited that she felt a considerable embarrassment and shame for what she recognized as a hereditary stigma.

This patient gave the history of affected relatives as mentioned above. A brother is said also to be affected but, so far as she knows, there are numerous other members apparently normal.

Examination showed the several pedunculated fibrous facial polyps of which this woman complained and, in addition, many more on the neck, trunk and limbs. They showed the two types commonly seen, namely, the pedunculated or sessile soft polyp and the flat bluish and compressible nodule resembling a haemangioma. These latter are reported histologically as "vascular neurofibroma." Pigmentation was prominent, and consisted for the most part of a freckle-like spotting with only a few larger patches.

No subcutaneous nerve-trunk tumour was observed nor was there any neurological abnormality; the ocular fundi were normal. There was a mild thoracic kyphoscoliosis.

- (b) The subcutaneous nerve-trunk swellings are usually less numerous and less conspicuous but are more prone to be a source of disability, if they are sensitive to touch, and subject to pressure in the course of normal activity. Spontaneous pain is unusual.

These lesions are typically spindle-shaped, and often demonstrably continuous with a normal or thickened nerve trunk. The size may commonly be as great as 8 or 10 centimetres in diameter, the consistency is a rubbery hardness. The nodules are freely mobile from side to side, but not along the length of the nerve of origin. The overlying skin is not adherent. Nerve dysfunction in the absence of compression or malignant change is rare, although Alexis Thomson described neuralgic pains, paraesthesiae and muscle twitchings.

- (c) Visceral lesions may be manifest in tumours of the abdominal or thoracic cavity, or of the stomach or other hollow viscus but these must be regarded as rarities. Nevertheless, at autopsy on a patient who has multiple neurofibromatosis visceral involvement is commonly seen.
- (d) The pigmentation appears to bear no relation in distribution to the fibromata. It may be no more than normally apparent on an average individual, or be the most obvious feature. The *café au lait* patches of pigmentation are the most characteristic type; the pigment is melanin, and the name, of course, derived

from the colour of these irregular, but well-defined, marks. The skin in the pigmented area is of normal texture, not elevated, and the patches vary in size up to ten or twelve centimetres in diameter (Fig. VI).

Very commonly there is, in addition, a more profuse scattered macular pigmentation, especially on the trunk, which may be diffusely covered with brown freckle-like spots, one or two millimetres in diameter (Figs. VI and VII).



FIG. VII. A *café au lait* patch, and sessile neurofibromata of the vascular and non-vascular types are seen.

- (e) The plexiform neurofibroma is much rarer. It may occur alone, or in the subject of a generalized neurofibromatosis. The most common site for these lesions is the face and neck, where they may produce unsightly masses and pendulous folds.

The condition is a localized and irregular fibromatous enlargement of nerve fibres, usually in the subcutaneous tissue, and produces a tumour which is soft and flabby in consistency, and the palpable thickened nerves are said to give it the sensation of a bag of worms. More accurate, perhaps, is the similarity to the

feeling of a pendulous breast in an emaciated old woman (Aird, 1949).

The histological lesion is essentially the same as in the generalized fibromatosis, and indeed the two forms often occur together, the plexiform lesions arising, apparently, from one of the nodular lesions.

- (f) Elephantiasis produced by neurofibromatosis may be regarded as a more advanced and diffuse degree of the one essential process. There is a massive enlargement of the soft tissues of the part, due to fibromatous overgrowth in the skin and subcutaneous tissues (Fig. VIII).



FIG. VIII. Neurofibromatous enlargement of the thumb and palm.

Usually a limb is affected; there may be thickening of the scalp or facial tissues, tongue or lips, producing wrinkled, pendulous folds. Fig. IX shows an early lesion of the lobule of the ear producing enlargement. The weight of the tissue alone may be a considerable handicap, and the wrinkled, elephantoid skin is subject to irritation and excoriation, being difficult to keep clean.

According to Holt and Wright (1948), actual increase in the size of bones in the affected part may occur. This is due to the neurofibromatous tissue causing elevation of the periosteum, with subperiosteal new bone formation. Either epiphyseal stimulation with increase in length or premature fusion with arrest in growth may be seen. Often the bones in relation to the elephantiasic tissue are slender and deformed, as if by compression.





FIG. IX. Neurofibroma producing enlargement of the ear-lobe.

#### ASSOCIATED LESIONS

A great variety of associated lesions has been noted in this disease, some of them causally related, some without other than chance relation, so far as can be seen. Skeletal changes are not uncommon.

##### (i) *Kyphoscoliosis*

This is the commonest skeletal defect in the disease, and may be of mild or severe degree. According to Holt and Wright (1948) it may be due to—

1. Disparity of the limbs, the result of the disease.
2. Anomalous vertebral development.
3. Erosive vertebral defect (from contiguous neurofibroma).

It must be confessed that cases are seen where it is difficult to incriminate any of these factors.

##### (ii) *Rarefactions*

Diffuse or localized rarefactions, with the appearance of subperiosteal or intra-osseous cyst formation is one of the commonest features. Shallow, pond-like depressions in the cortex and compressed, deformed bones

are produced by the indenting effect of juxta- or sub-periosteal neurofibromatous tissue.

These osseous lesions are presumed to originate from periosteal nerves; the evidence for the presence of nerves within normal bones is as yet conflicting.

Secondary deformities may occur, and the subjects of the disease are occasionally dwarf-like and grotesque. Fractures of the involved long bones are prone to be followed by non-union, but histological proof of the presence of neurofibromatous tissue at the site is not often available.

##### (iii) *Developmental abnormalities*

All authorities agree that the incidence of other developmental defects is increased. Club-foot is not uncommon.

##### *Case 3*

Some of these features are illustrated in the record of a patient, a man aged 46, who died eight weeks after admission with carcinoma of the head of the pancreas.

He was a dwarf and had been small for as long as he could remember. There was no recognizable family history of neurofibromatosis. His skin showed soft sessile nodules ranging from 2 to 10 millimetres in size, scattered over the limbs, trunk and face. There were pigmented patches from pinhead size up to 5 centimetres in diameter on the trunk. No deeper tumours were noted. There was a scoliosis and a deformity of the right foot. There was a mild bilateral exophthalmos, and the ocular fundi showed a peculiar tigroid distribution of the choroidal pigment. No other neurological abnormality was present.

There was jaundice, wasting and the expected concomitants of the pancreatic neoplasm.

Radiographs confirmed the gross thoraco-lumbar scoliosis with marked rotation of the vertebral bodies. There was a coxa valga, with marked coarsening of the trabeculae in the proximal third of the femoral shafts.

The right foot showed chronic degenerative changes in ankle and mid-tarsal joints. All the metatarsals were remarkably slender, and there was marked hallux valgus and pes cavus. The os calcis, astragalus and cuboid bones were fused into one amorphous mass.

##### (iv) *Orbital lesions*

Orbital roof defects have been recorded, usually with exophthalmos which may be pulsating (transmitted cerebral pulsation). In some of these, neurofibromatous infiltration of the upper lids and retrobulbar tissues is present, and adequately explains both the

bony defect and the exophthalmos. In others, an extensive bony defect without other local signs of neurofibroma is seen.

(v) *Limb lesions*

Congenital bowing of the tibia, leading to fracture and pseudarthrosis, has a now generally accepted association with multiple neurofibromatosis (Vieta and Pack, 1951) which may become apparent only when the typical lesions appear at puberty or later.

#### NEUROLOGICAL LESIONS

It might be expected that a disease of the supporting tissues would have numerous lesions of the central nervous system, but they are not common.

- (a) Auditory nerve tumours (described usually as acoustic neuromas, or neurilemmomas) have a recognized association with neurofibromatosis, and are very often bilateral. The stigmata of the disease should be sought in all cases of auditory nerve tumours.
- (b) Meningiomas, sometimes multiple, have been reported in association with neurofibromatosis, but these tumours are rarely present.
- (c) Extradural and intervertebral neurofibromas are a well-recognized cause of compression of the spinal cord and spinal nerve and produce signs and symptoms according to their site.
- (d) Deafness and optic atrophy occur, sometimes demonstrably due to fibrous thickening of the nerves. Choroidal pigmentary abnormalities and retinal defects have been seen.

The incidence of feeble-mindedness is greater than in a comparable series of other cases (Preiser and Davenport, 1918).

#### MALIGNANT TUMOURS IN NEUROFIBROMATOSIS

The frequency of this complication is usually stated as 10 to 15 per cent. of cases but one would think that if all the asymptomatic and the mild forms of the disease are considered, this is much too high a figure.

It is interesting that malignant change in the cutaneous lesions is rare, although there is no obvious reason for this. The malignant

neurofibroma is usually found in a subcutaneous or deeper nerve, commonly of an extremity, but sometimes intra-abdominal or thoracic (Hermann, 1950). A major nerve trunk such as the sciatic is not uncommonly affected (Case 4).

#### Case 4

A man aged 35 years felt a sudden pain in his left leg during exertion, and afterwards noticed a lump behind the knee. Later another appeared behind the right knee. This lump was subsequently excised in another hospital, but it returned eleven months later.

He was then seen at this hospital, when he gave the history that the mass behind the left thigh had been increasing in size and was painful.

He was found to have a generalized neurofibromatosis, without any family history of the condition. There was a healed scar behind the right knee. In the posterior compartment of the left thigh and extending downwards to the knee was a large, hard, adherent mass. The muscles of the lower leg and foot were weak and wasted, but there was no other neurological abnormality. There was a lower dorsal scoliosis to the right side.

Radiographs revealed no bony lesion in the thigh and no erosive vertebral defect.

With a diagnosis of malignant neurofibroma, a high amputation was performed. The examination of the specimen revealed a spindle-cell sarcoma in close association with the sciatic nerve.

This patient was lost to follow-up until four years later when he died in another hospital. The autopsy findings were reported as "bronchopneumonia, cachexia, neurofibromatosis."



FIG. X. Low power view of the malignant neurofibroma removed from Case 4, showing the very cellular fasciculated arrangement. (x 33)

The onset of malignancy may be insidious, but is usually indicated by the appearance of pain and an increase in the size of a previously inoffensive lump in the limb of an

adult patient having multiple neurofibromata. Growth of the tumour is then rapid and it may become tender, hot, and of a large size. Paralysis or paraesthesiae may appear in the region of the affected nerve trunk.

According to Hosoi (1931) the malignant change is said to be prone to follow any surgical intervention, and this has been supported by other writers; but it would seem more likely that it is the associated change in symptoms which first initiates a demand for surgery.

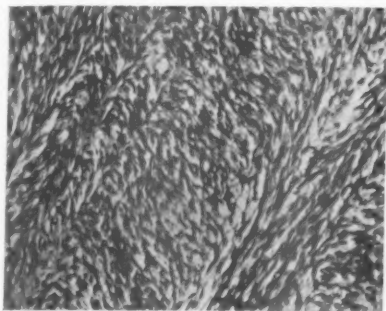


FIG. XI. Higher magnification of the tumour shown in Fig. X. (x 175)

The malignant tumours may at first appear well encapsulated, but local removal is prone to be followed by recurrence and microscopic spread along the affected nerve trunk for some distance beyond the microscopic limits can be demonstrated. At a later stage it may be difficult to establish the precise origin of a large sarcoma.

Histologically, the great change in appearance with the onset of malignancy can be seen from Figs. X and XI. There is a considerable increase in the number and nature of the large, elongated cells which are banded together in great masses and fasciculi, with sometimes evidence of the palisade effect which is an indication of their origin. Pleomorphism, giant cells, and mitoses are common.

The degree of malignancy would appear to be high. Metastases are, as a rule, late, and occur mainly in the lungs. The treatment of

choice is radical local excision, or amputation (Vieta and Pack, 1951).

### CONCLUSIONS

To the patient, multiple neurofibromatosis may be of no significance, may be a source of embarrassment or shame, or constitute a dangerous threat to life.

The treatment of the disease must, by its very nature, be palliative: the problems are, for the most part, mechanical. Nevertheless, it may exercise the dermatologist, neurologist or ophthalmologist, the orthopaedic, thoracic and plastic surgeons, and present to them all problems of diagnosis and treatment.

### ACKNOWLEDGEMENTS

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### REFERENCES

- AIRD, I. (1949), "A companion in Surgical Studies." Edinburgh, Livingstone, page 350.
- BEATTIE, J. M. and DICKSON, W. E. (1943), "Text-book of Pathology." London, Wm. Heinemann, Fourth Edition.
- HERRMAN, J. (1950), *Ann. Surg.*, vol. 131, page 205.
- HOLT, J. F. and WRIGHT, E. M. (1948), *Radiology*, vol. 51, page 612.
- HOSOI, K. (1931), *Arch. Surg.*, vol. 22, page 258.
- MASSON, P. (1932), *Amer. J. Path.*, vol. 8, pages 367, 389.
- PREISER, S. A. and DAVENPORT, C. B. (1918), *Amer. J. med. Sci.*, vol. 6, page 507.
- SHARPE, J. C. and YOUNG, R. H. (1937), *Arch. intern. Med.*, vol. 59, page 299.
- THOMSON, H. A. (1900), "On Neuromes and Neurofibromatosis." Edinburgh, Turnbull and Spears.
- VIETA, J. O. and PACK, G. T. (1951), *Amer. J. Surg.*, vol. 82, page 416.
- VON RECKLINGHAUSEN, K. (1882), "Über die multiplen fibrome der Haut und ihre Beziehung zu den multiplen neuromen." Berlin, A. Hirschwald.
- WEBER, F. P. (1909), *Brit. J. Derm. Syph.*, vol. 21, page 49.

## RENAL LEIOMYOMATA

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THE paucity of literature describing non-epithelial tumours of the kidney gives the impression that these growths are rare. That such is not the case is evident from the increasing number of reports of the condition especially within the last two decades.

The first muscle tumour of the kidney was recorded in 1901 by Lartigan and Larkin, two cases of benign subcapsular nodules of smooth muscle being described.

No further report can be traced until 1929 when a single tumour measuring 13.5 cm. in diameter was reported by Bugbee. In the same year Berry discovered 2 further cases, the tumour in one being benign, that in the other being regarded as malignant.



FIG. 1. Photograph of a benign leiomyoma in an ectopic kidney (Case 6). Note its peripheral position.

Another malignant growth was reported in 1932 by Crosbie and Pinkerton, and in the following year an additional leiomyosarcoma was described by Swan (1933).

It was also in 1933 that Cooke gave an account of a leiomyosarcoma of the kidney

with extensive metastases — the first record of metastases associated with this growth. Independent reviews of the literature were conducted in 1937 by Mintz and Patch who collected 6 and 10 cases respectively. Later work by Weisel, Dockerty and Priestley (1943) revealed a further 3 malignant tumours. A comprehensive survey of all reported cases was published by Kretschmer in 1952, the total number of malignant growths, including his own case, being 15.

Kerr in 1954 added one further example in which removal of a gastric leiomyosarcoma one year previously, preceded the discovery of a renal tumour of similar histology.

In this present paper a series of muscle tumours of the kidney comprising 7 cases of benign leiomyomata and 4 examples of malignant muscle tumours, is given.

### CASE DETAILS

#### (A) *The benign leiomyomata*

This group consists of 7 cases, 5 of which gave no symptoms of renal disease, all being found incidentally at autopsy. The 2 remaining patients presented with a mass within the abdomen.

##### *Case 1*

Male, aged 57 years, died of exfoliative dermatitis. No renal symptoms had occurred.

##### *Case 2*

Female, aged 65 years, cause of death not known. No renal symptoms had occurred.

##### *Case 3*

Female, aged 60 years, died of carcinoma of sigmoid colon. No renal symptoms had occurred.

##### *Case 4*

Female, aged 53 years, died of carcinoma of stomach. No renal symptoms had occurred.

##### *Case 5*

Male, aged 25 years, died of cerebral tumour. No renal symptoms had occurred.

**Case 6**

Female, aged 47 years, for 4 weeks had been conscious of a lump within the abdomen, this being associated with a dull ache.

Examination confirmed a mass in the left loin, the mass being considered to be kidney. A preliminary straight X-ray revealed 2 normal kidney shadows, so pyelography was not carried out. Laparotomy however revealed an ectopic third kidney with an attached encapsulated mass 8 cm. in diameter (Fig. 1) which was proved histologically to be a leiomyoma. This patient is still alive and well six years after operation.



FIG. II. Photograph of a large leiomyosarcoma showing the presence of many cystic spaces (Case 10) and the absence of gross haemorrhage. Again it is peripherally placed.

**Case 7**

Female, aged 68 years. This history here was of periodic left-sided abdominal pain for nine months. Three months prior to admission to hospital a swelling was noticed in the left loin. At no stage were there any urinary symptoms. Examination revealed a palpable mass in the left flank and pyelography showed delayed excretion of dye with distortion of the calyces of the left kidney. At operation an encapsulated tumour weighing 43 ozs. was found. The histological picture of this tumour was of well differentiated smooth muscle, though in one section sufficient atypical cells were seen to justify a guarded prognosis. No records of post-operative survival are available.

There now remains a group of 4 examples of the malignant tumour. These tumours are usually larger, when detected, than the benign growths, probably due to the more rapid rate of growth. Symptoms of renal disturbance are not common however, only one patient presenting with haematuria.

**(B) The leiomyosarcomata****Case 8**

Female, aged 35 years, had been perfectly well until nine days before admission to hospital when she developed profuse painless haematuria. On

examination no mass was demonstrable. The haemoglobin was 75 per cent., blood was apparent in the urine, and pyelography showed gross deformation of the calyces of the right kidney. Nephrectomy was performed, an encapsulated tumour measuring 8 cm. in diameter being removed.

The histological report stated that the tumour appeared of slow growth with probably a relatively low malignancy. No records of survival are obtainable.

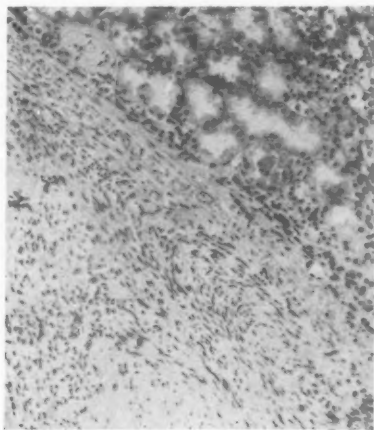


FIG. III. Photomicrograph of a benign growth (Case 4) showing parallel muscle bundles. Inflammatory cells are scattered throughout. (x 90)

**Case 9**

Female, aged 76 years, had a history of past intermittent diarrhoea for twenty years. She now presented with nausea, anorexia, and a lump in the right flank, the present symptoms being of three weeks' duration. The patient had lost one stone in weight and complained of no urinary symptoms. On examination a large mass was palpable in the right flank, but was not palpable from the loin. A provisional diagnosis of carcinoma of the ascending colon was made and, without further investigation, operation was undertaken. The growth was found to arise from the lower pole of the right kidney spreading beyond the confines of the capsule, involving the vascular supply of the ascending colon and necessitating removal of the ascending colon and caecum, with the kidney. The histological picture in this case was one of extreme anaplasia (Figs. VII and VIII). Ten months after operation the patient is very well and has gained 2 stone in weight.

**Case 10**

Female, aged 80 years, one month prior to presentation had a three-day period of left-sided abdominal pain and constipation, both being relieved by an enema. On straight X-ray the left renal outline could not be identified and an intravenous pyelogram demonstrated non-filling of the upper



major calyx on the left side. A large semi-fixed mass was palpable in the left loin, but at this stage, although a presumptive diagnosis of neoplasia was made, surgery was not undertaken. Ten months later, as the mass had enlarged considerably, still without producing any symptom of renal disease, it was removed and found to be 24 cm. in diameter. Histologically this tumour was undoubtedly malignant (Fig. VI). At the present moment, three months after operation, the patient is well and has gained 2 stone in weight.

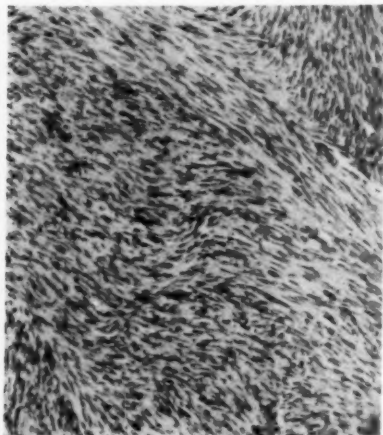


FIG. IV. Photomicrograph of a benign growth (Case 1) showing well-developed muscle tissue. (x 90)

### Case 11

Female, aged 64 years, had complained of increasing tiredness and loss of weight during the preceding twelve months and had experienced no urinary troubles. On examination a large, firm, rounded mass 20 cm. x 24 cm. was palpable in the left loin, and pyelography showed a non-functioning kidney. The haemoglobin was 55 per cent. with no history of blood loss. Left nephrectomy was performed and a mass 30 cm. in diameter was found with only a small amount of normal renal tissue. Death of the patient ensued seven months later, at which time there was extensive local recurrence in the loin. Post-mortem examination was not made.

### PATHOLOGICAL FEATURES

The benign muscle tumour is firm, pearly-grey in colour, usually small and varying in diameter from 1.3 cm. In two instances the tumour was larger and one undoubtedly benign growth measured 8 cm. in diameter. The other large tumour, weighing 43 oz. (Case 7) and here classified as benign had some features to warrant a suspicion of malignancy.

The malignant tumour at the time of diagnosis was much larger, the average diameter being approximately 20 cm. and appeared more cellular than its benign counterpart. Both tumours appeared well encapsulated.

Macroscopically on gross section, muscle tumours in no way resemble the variegated haemorrhagic tumour so typical of carcinoma of the kidney.

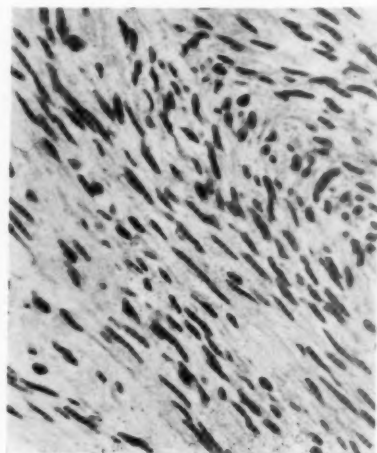


FIG. V. Photomicrograph (Case 8) demonstrating well-differentiated muscle tissue but with also a few hyperchromatic nuclei. (x 360)

The small benign tumours were all situated immediately subjacent to the capsule. Of the malignant group, the tumour in three instances was peripherally placed, the site of the fourth in relation to the renal parenchyma being uncertain. Two of the malignant examples were examined carefully and in each case the tumour was found to be contained within a fascial capsule common to the tumour and also to the kidney, but the growth apparently had no other connection with the renal parenchyma (Fig. II). Cystic spaces were scattered throughout one specimen (Fig. II) and in another necrosis was prominent. Haemorrhage however was rare and when present was never massive as it often is in carcinoma of the kidney.

Histologically the structure of the growth varies from the well-differentiated benign tissue to an extreme degree of anaplasia.

The benign lesion usually shows no delineating capsule between growth and renal substance unless the tumour has attained a considerable size. It is composed of typical smooth muscle cells which are elongated or spindle-shaped cells arranged in interlacing bundles, exhibiting elongated or oval nuclei, rounded or blunt at the ends, and possessing a distinctly granular appearance. The cytoplasm is longitudinally fibrillated and of moderate amount (Figs. III and IV).

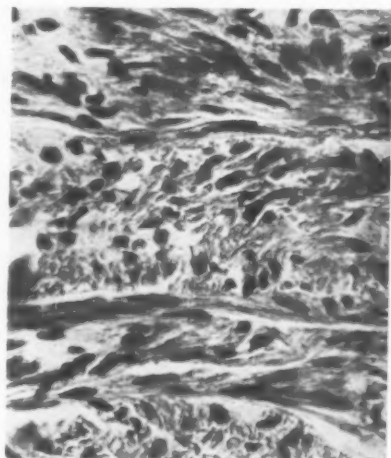


FIG. VI. Photomicrograph (Case 11) of a tumour of undoubted malignancy. Muscle bundles however are still apparent. (x 360)

The transition to the malignant phase would appear to be insidious, for mitotic figures are not a common feature unless the tumour is frankly malignant; the presence of clumped hyperchromatic nuclei in an otherwise well-differentiated growth warrants a suspicion of malignancy (Fig. V). The obviously malignant growth is very cellular, with scant intercellular substance. Muscle bundles run parallel to each other, interlace, and run in different directions, thus still resembling superficially the benign tumour, but the individual cells vary considerably in size and staining properties, the pattern being obviously malignant (Fig. VI). Since differentiation in the benign lesion and the mature tumour is well developed, there is then no problem in determining the nature

of the tumour; on the other hand extremes of anaplasia occur resulting in difficulty in determining the histology of the primitive tumour. Figs. VII and VIII illustrate the histology of an anaplastic tumour (Case 9). However, in other portions of the same section, fairly well-differentiated muscle tissue can be identified, and in this way the diagnosis can be made in what would otherwise be a difficult case.

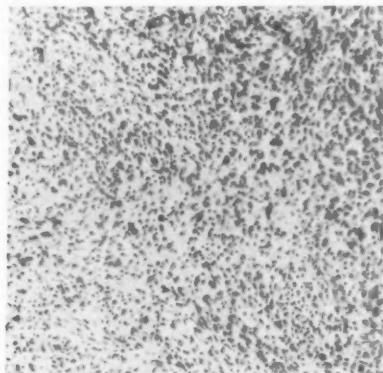


FIG. VII. Photomicrograph (Case 9) of an anaplastic tumour. There is no resemblance to muscle tissue in this particular field. (x 90)

#### DISCUSSION

This series of 11 cases of renal muscle tumours, seven benign and four malignant, is reported so that emphasis is placed on the existence of non-epithelial tumours of the kidney, an increasing number of which are being described.

Thus of the eleven cases presented, only 2 occurred in the male sex, 9 being in females, whilst haematuria was a symptom in only one instance. These findings are very different from the behaviour of carcinomata of the kidney where the incidence is mainly in the male sex, and where haematuria is the commonest single presenting symptom. It should be noted, however, that of the malignant muscle tumours previously recorded, the sex distribution was of almost equal distribution and haematuria was a feature in 50 per cent. of cases.

In this series the small benign lesion ranging in size from 1.3 cm. was observed as an incidental finding at autopsy in 5 cases. The other 2 benign tumours were of larger dimensions and presented with a mass, as did three of the four malignant growths.

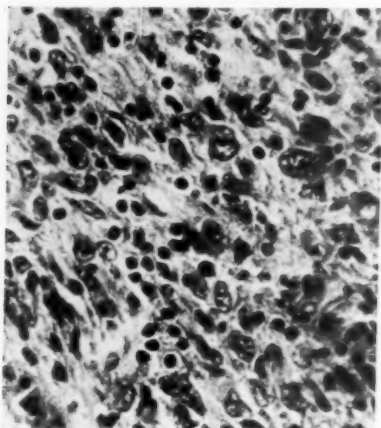


FIG. VIII. Photomicrograph (Case 9) of the same tumour as in Fig. VII. The nuclear pattern is bizarre. (x 350)

The small cortical tumour of 1.3 cm. in diameter was never detected clinically and it would be expected that a mass must be of considerable size before it is detected by the palpating hand. Haematuria was a feature in one case only, and therefore the clinical appreciation of the lesion in this series depended on the sole feature of the size of the tumour. It therefore follows, in view of the larger size and more rapid growth of the malignant tumour, that the benign lesion will continue to be unsuspected until revealed at autopsy. Theoretically the growth may arise from the smooth muscle existing in the capsule, around blood vessels, or in the pelvis. In the 10 cases where the site is recorded the growth was always cortically placed, no matter how large the tumour. It thus seems that as these tumours enlarge they do not tend to encroach on the pelvis as do carcinomata of the kidney, but maintain their peripheral situation. This may be related to the fact that the large muscle tumour is almost invariably invested by a well-developed fascial capsule.

The maintenance of the peripheral position can be correlated with the presenting features. Carcinomata of the kidney notoriously spread at the expense of the renal parenchyma, and may attain a considerable size before the renal dimensions are increased. The peripheral myosarcomata however enlarge not at the expense of the kidney but more or less apart from the normal sized kidney and would thus be more obvious to the clinician.

Haematuria was present in one case only in this series, the site of the tumour in this case being uncertain. In all cases where the tumour was known to be peripheral, haematuria did not occur, and it seems probable that as in carcinomata of the kidney haemorrhage into urinary passages seldom occurs unless the pelvis of the kidney is invaded by tumour tissue. In the one case, with blood in the urine, it is possible that the tumour arose from the muscle of the lining of the pelvis, but on the other hand, it might have been due to raised venous pressure caused by the tumour obstructing venous reflux from the vessels in or around the pelvis. It follows that symptoms of renal disease in this particular tumour do not become manifest until the tumour has attained a considerable size, and surgical ablation of the lesion will then not take place until the tumour has been present for a considerable period of time. The association of metastases with the lesion is apparently uncommon, however, and thus surgical cure seems feasible, the prognosis being far better than for carcinomata of the kidney. In view of the small number of cases, and the unpleasant fact that the autopsy room reveals many metastases which were not suspected clinically, it is of paramount importance that, when death ensues, a post-mortem examination is conducted on all cases. Not until then will the natural history of this disease and the value of surgery in its relief, be fully assessed.

#### ACKNOWLEDGEMENTS

I am indebted to the Honorary Medical Staff of the Alfred Hospital, Melbourne, and to Mr. Robert Officer, Mr. J. B. Somerset and Dr. M. B. Wanliss for access to records of their patients. I have to thank Mr. J. C. Dick, Medical Superintendent of the Ballarat Base Hospital, and Dr. R. J. Riddell, Pathologist

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## REFERENCES

- BERRY, W. E. (1929), *Canad. med. Ass. J.*, vol. 20, page 280.
- BUCBEE, H. G. (1929), *J. Urol.*, vol. 21, pages 363-369.
- COOKE, W. E. (1933), *J. Path. Bact.*, vol. 37, page 157.
- CROSBIE, A. N. and PINKERTON, H. (1932), *J. Urol.*, vol. 27, page 27.
- KERR, J. A. (1954), *Brit. J. Surg.*, vol. 41, page 478.
- KRETCHMER, H. L. (1952), *J. Urol.*, vol. 68, page 36.
- LARTIGAN, A. J. and LARKIN, J. H. (1901), *J. med. Res.*, vol. 1, page 25.
- MINTZ, E. R. (1937), *Ann. Surg.*, vol. 105, page 521.
- PATCH, F. S. (1937), *Brit. J. Urol.*, vol. 9, page 345.
- SWAN, R. H. J. (1933), *Brit. med. J.*, vol. 1, page 606.
- WEISEL, W., DOCKERTY, M. G. and PRIESTLEY, J. T. (1943), *J. Urol.*, vol. 50, page 567.

## PRIMARY ONE-STAGE PROCTOCOLECTOMY IN ULCERATIVE COLITIS

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**A**LTHOUGH non-operative measures can control many cases of ulcerative colitis, it is generally accepted that a number requires surgical treatment. When surgery is indicated it is agreed that the colon must be removed, and most feel that the rectum should be excised as well.

The orthodox method of performing this excision entails two or three separate operations. At the first, an ileostomy is constructed; at the second, the colon is removed; and at the third, an abdomino-perineal excision of the rectum completes the removal of the large bowel.

As experience in the surgical treatment of this disease has developed, and with improvement in conditions under which the operation is performed, the surgical approach has become bolder and many now advocate and practise excision of the colon at the time of ileostomy, even in the seriously ill patient. Indeed, it is our opinion, that the more desperate the patient's condition, the more urgent is his need for colectomy. Unless involvement is minimal, the rectum is subsequently removed.

In this series of cases, the whole procedure of ileostomy, colectomy, and excision of the rectum has been carried out at the one operation.

### CASE HISTORIES

#### *Case 1*

Mrs. J.R., aged 31 years, was first seen in August, 1954, complaining of recurrent diarrhoea over the past four years; in the last year her symptoms had been persistent. Motions averaged five daily; they were unformed, contained much mucus, but only on occasions had she observed the presence of blood. There was no loss of weight and no symptoms suggestive of toxæmia.

Clinically, she appeared to be a mild case, but rectal examination showed a thick, fibrous stricture commencing five centimetres from the anus, and through which it was impossible to pass a sigmoidoscope. The mucosa was inflamed and oedematous. A barium clysm showed involvement of the whole colon and terminal ileum.

In view of the gross changes in the large bowel and the presence of the stricture which was thought to be possibly malignant (later proved otherwise), operation was advised. This was performed on the 5th October, 1954. The rectum was first removed, and as her condition was quite steady, her colon was then excised and ileostomy constructed through the right rectus muscle. A split skin graft was applied to the ileostomy (Fig. 1).

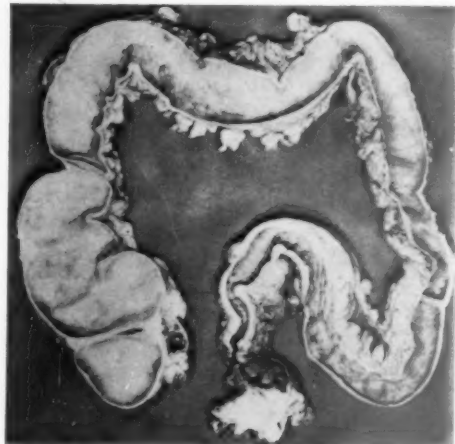


FIG. 1. Case 1: The colon and rectum after removal. There is a stricture in the upper third of the rectum. The remainder of the large bowel is contracted, particularly on the left side. There are numerous small ulcers in the rectum and sigmoid colon, and above this level the mucosa shows microscopic evidence of atrophic changes.

Her convalescence was uneventful and she was discharged home on the 20th post-operative day. Apart from developing an ileostomy stenosis, she has remained very well.

#### *Case 2*

Mrs. A.L., aged 66 years. This patient had suffered from diarrhoea for 25 years; she had not passed a formed stool during the whole of this time. The average number of bowel actions was six daily, and on occasions she observed blood. In the last six months she had lost one stone in weight, her appetite had fallen away, and her energy had lessened considerably.

On examination, she appeared sick, anaemic, and had obviously lost much weight. The liver was enlarged two centimetres. Sigmoidoscopy revealed



gross ulceration and loss of distensibility within the rectum and lower sigmoid colon. A barium clyisma showed advanced ulcerative colitis; a narrowing just below the splenic flexure almost certainly represented a carcinoma.

Operation was required urgently, but it was felt that her general condition was so poor that only a local resection would be feasible. On the 19th December, 1954, laparotomy was performed; a small, annular carcinoma was situated just below the splenic flexure. No macroscopic secondary deposits could be found. The inferior mesenteric artery was ligated at the aortic origin and the inferior mesenteric vein secured at the lower border of the pancreas. The left side of the colon was then freed. The condition of the patient was quite steady, and accordingly the dissection of the colon was continued around to the caecum and down to the recto-sigmoid junction; an ileostomy was constructed through the right rectus sheath and muscle. Her condition continued to be excellent and so the excision of the large bowel was completed with removal of the rectum. A skin graft was applied to the ileostomy (Fig. II).

### Case 3

Mr. McD., aged 48 years. This patient had suffered from intermittent attacks of diarrhoea, with bleeding, for more than twenty years. He suffered a severe exacerbation in November, 1953; at this time, sigmoidoscopy revealed an ulcerative proctocolitis, but a barium clyisma showed the colon, above the recto-sigmoid junction, to be normal in appearance. Over the next few months, the diarrhoea persisted, but was not unduly troublesome until August, 1954, when there was a further severe exacerbation. His bowels acted at half-hourly intervals; his appetite fell away and the thought of food made him nauseated. His weight dropped quickly and he became too weak to continue with any work. Sigmoidoscopic examination, in September, 1954, revealed a similar appearance as was found a year previously, except for the appearance of pseudo-polypi at eighteen centimetres. A second barium clyisma indicated extension upwards to involve the whole of the sigmoid colon.

He showed no favourable response to medical treatment, and it was decided that operation must

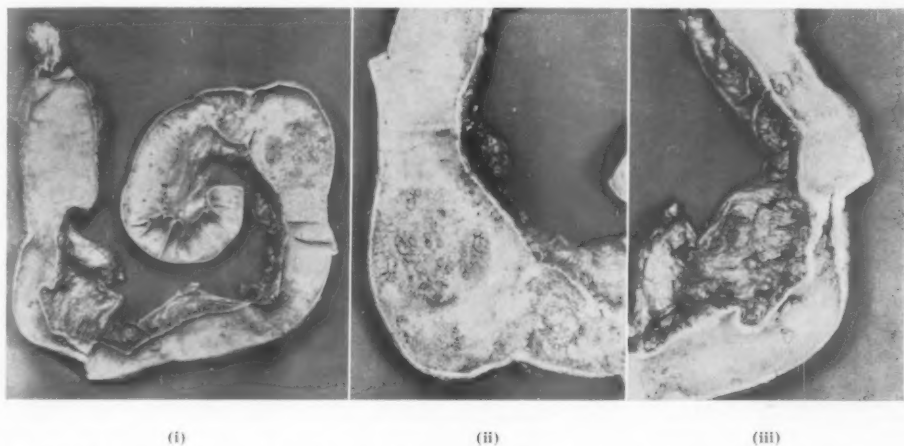


FIG. II. Case 2: (i) The colon and rectum after removal. There is a neoplasm of the descending colon. The remainder of the large bowel is contracted with small areas of ulceration and pseudo-polypi. The pedicle of the tumour is the inferior mesenteric artery, ligated at the origin from the aorta.

(ii) Close-up view of the caecum, showing the pseudo-polypi.

(iii) Close-up view of the carcinoma in the descending colon. Note the presence of other polypi and the pedicle, which was ligated just below the third part of the duodenum on the aorta.

Her immediate convalescence was smooth. On the 10th post-operative day she had a melaena stool, and this necessitated a blood transfusion; the source of the bleeding was not definitely determined but was thought to be a peptic ulcer. She left hospital at the end of the fourth week after operation. She has remained well, except for stenosis of the ileostomy; this has required operative relief. Six months after operation there was no sign of recurrence of the tumour and she had gained one and half stone in weight.

be done. In view of the gross involvement of the rectum and sigmoid colon, together with the presence of pseudo-polypi, and, because of the radiological freedom of the more proximal large bowel, it was felt that operation should commence with removal of the rectum and sigmoid colon. On the 11th October, 1954, laparotomy was performed; the presence of colitis to the hepatic flexure was observed. An abdomino-perineal excision of the rectum was performed, and, as his condition was satisfactory, the colectomy was completed, and an ileostomy constructed through the right rectus muscle. A split skin graft was applied to the ileostomy (Fig. III).

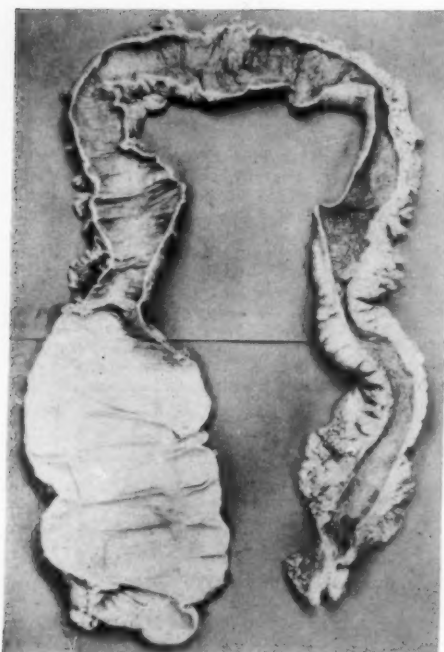


FIG. III. Case 3: The colon and rectum after excision. The caecum and neighbouring ascending colon are little affected by the colitis. The left half of the colon and the rectum are involved to a severe degree with ulceration and pseudo-polypi.

His convalescence was uneventful and he was discharged home from hospital on the 24th October, 1954. A week later the ileostomy stoma was enlarged a little; he returned to work in the sixth week after operation, and has been in excellent health since.

#### Case 4

Mr. L.D., aged 44 years. This patient had been aware of some looseness of the bowels, with the passage of excess mucus, for about ten years. Two years prior to the present illness, he had observed some blood in the motions, and, one year previously, he had had an acute bout of diarrhoea, lasting two weeks, but from which he had made a spontaneous recovery. Over the past two months, he had a further exacerbation, which was getting worse. He was having twelve motions a day, was losing weight rapidly, and complained of persistent anorexia and nausea.

He appeared very sick, and this was accentuated by superficial skin sores scattered around his mouth. Sigmoidoscopy revealed advanced ulcerative proctocolitis and a barium clysm showed a typical involvement of the rectum and sigmoid colon, but the remainder of the colon appeared normal, radiologically.

He was treated medically for three weeks, in which time his temperature was persistently elevated. His condition showed no material improvement, and operation was advised as a life-saving procedure. In view of the gross involvement of the rectum and distal colon, it was felt that this should be removed first. On the 23rd December, 1954, operation was performed; the colon was found to be inflamed from the middle of the transverse colon, to the rectum; proximal to this is appeared normal. An abdomino-perineal excision of the rectum was performed, and as his condition was quite satisfactory, the remainder of the colon was removed (Table 1). An ileostomy was constructed through the right rectus muscle, and a skin graft applied (Fig. IV).

His convalescence appeared to progress uneventfully, and was associated with an immediate and remarkable improvement in his feeling of well-being. However, on the 4th January, 1955, whilst having a bath, he collapsed and died within a few minutes. Autopsy revealed a massive pulmonary embolism; the peritoneal cavity was clear.

TABLE I

*Information obtained from the anaesthetist's record of the operation performed on Mr. L.D. (Case 4).*

Mr. L.D.—OPERATION, 23rd DECEMBER, 1954.			
Time	Pulse Rate/Minute	Blood Pressure Millimetres of Mercury— Systolic/Diastolic	Stage of Operation
1.50 p.m.	110	120/80	Immediately prior to induction
2.19 p.m.	88	110/85	Operation commenced
3.55 p.m.	100	115/90	Peritoneum closed
4.20 p.m.	100	120/80	Just before leaving theatre

## Case 5

Mrs. N.D.G., aged 41 years. She had been in excellent health until two weeks before her admission to hospital. Almost simultaneously, she developed a persistent diarrhoea, and a discharging perineum. She soon became quite incontinent. The aecal discharge was liquid, profuse, and heavily stained with blood. She was anorexic and vomited on occasions.



(i)

FIG. IV. Case 4: (i) The colon and rectum after excision. There is minimal involvement of the caecum and neighbouring ascending colon. The very severe ulceration in the rectum and sigmoid colon is visible, whilst the descending colon and transverse colon are involved to a lesser degree.



(ii)

(ii) A close-up view of the sigmoid colon to demonstrate the very severe and extensive ulceration of the mucosa.

She was very apprehensive and confused. Her temperature was 103°F and remained elevated although irregular in pattern. There were numerous fistula openings around the perineum; there was a large, ragged recto-vaginal fistula; the anal sphincters appeared to have been destroyed. Sigmoidoscopy revealed large, but superficial, ulcers scattered in the rectum and sigmoid colon, with the intervening mucous membrane oedematous and inflamed. A barium clysis was done with difficulty, but it showed advanced colitis of the descending and sigmoid colons; proximal to these portions, the large bowel appeared normal.

After admission to hospital, there was no material change in her condition until a severe haemorrhage necessitated the rapid administration of several pints of blood; bleeding continued, and it was decided that operation should be done urgently, and should begin with the removal of the rectum. On the 19th October, 1954, a laparotomy was performed; the colon was involved in the inflammatory process from rectum to the mid-transverse colon. The rectum was excised, and as her condition was quite satisfactory, the remainder of the colon was removed and an ileostomy was constructed through the right rectus muscle (Fig. V).

Her convalescence was uneventful, and, apart from mild wound sepsis, the wounds healed quickly. She returned to her home in the country five weeks after the operation. She has remained well since then.

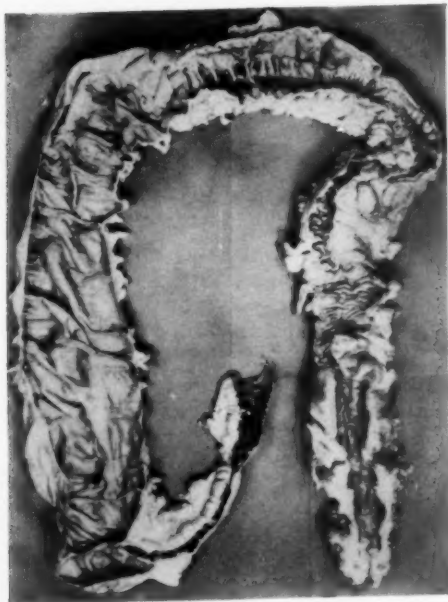
## Case 6

Mr. H.F., aged 42 years. He was admitted to the Royal Melbourne Hospital on the 15th December, 1954, complaining of diarrhoea, and the passage of blood over the past two weeks. He had never had

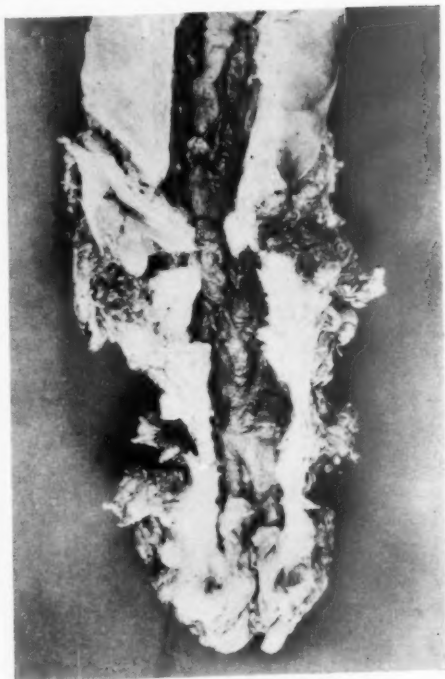
any previous trouble with his bowels, but now was passing up to twelve motions daily; the motions were quite unformed and consisted of mucus, blood, and faecal material. His appetite had fallen away, and he vomited on occasions. Sigmoidoscopy revealed an ulcerative proctocolitis, and barium enema examination disclosed involvement of the colon to the middle of the transverse colon, but proximal to this, the appearances were almost normal. He declined further hospital treatment and returned home, but was readmitted on the 17th January, 1955, in a grave condition. Just before admission, and after a bout of vomiting, he had become drowsy and semi-stuporose.

Dehydration was corrected by intravenous therapy; and the haemoglobin (66 per cent. 9.7 G. per 100 cc.) and serum protein (5.3 G., 13 milliequivalents per litre) levels partially restored by transfusion of whole blood. There was a temporary improvement, but this was not maintained and he appeared to be on the verge of a serious relapse when operation was done on the 21st January, 1955. An ileostomy, through the right rectus muscle, was first constructed; the right colon and then the left colon were freed, and, as his condition remained satisfactory, the rectum was finally removed (Table 2). The ileostomy stoma was covered with a split-thickness skin graft (Fig. VI).

Following this operation, his condition improved, at first slowly, and then rapidly; his convalescence was relatively uneventful, and he was discharged home on the 11th February, 1955.



(i)



(ii)



(iii)



(iv)

TABLE 2

*Information obtained from the anaesthetist's record of the operation performed on Mr. H.F. (Case 6).*

Mr. H.F.—OPERATION, 21st JANUARY, 1955.			
Time	Pulse Rate/Minute	Blood Pressure Millimetres of Mercury — Systolic/Diastolic	Stage of Operation
8.30 a.m.	72	110/70	Immediately prior to induction
9.00 a.m.	80	105/70	Operation commenced
11.15 a.m.	90	100/80	Peritoneum closed
11.25 a.m.	90	100/80	Just before leaving theatre

#### DISCUSSION ON THE CASES OF THIS SERIES

Two of these cases were suffering from chronic ulcerative colitis. Mrs. J.R. (Case 1) had a stricture in the rectum associated with advanced colitis; Mrs. A.L. (Case 2) had a long history of ulcerative colitis, but in recent months her symptoms changed for the worse, and investigation revealed a carcinoma of the descending colon. In both patients operation was done as an elective procedure; neither were regarded as serious risks, although their general condition (particularly that of Mrs. A.L.) was poor. In Mrs. J.R. the rectum was first removed, and then, as her condition was good, the colon was excised. In Mrs. A.L. the colon was first removed, and finally the rectum.

Two cases might be classified as sub-acute or chronic relapsing ulcerative colitis. Mr. McD. (Case 3) had had intermittent attacks of diarrhoea for twenty years, and Mr. L.D. (Case 4) had been similarly troubled for ten years. Relapses brought both patients to

hospital in a serious condition and in neither did medical treatment seem likely to bring a remission. In both there was clinical evidence of toxæmia; and in both, successive radiological examinations revealed upward spread in the colon. In both patients, operation was regarded as a life-saving measure; excision commenced with the rectum and finished with colectomy and ileostomy.

Two cases were regarded as fulminating ulcerative colitis. Mrs. N.D.G. (Case 5) had been well until a fortnight before admission, and was gravely ill with a high temperature, profuse diarrhoea and severe perineal sepsis. Mr. H. F. (Case 6) had also been free of trouble until a fortnight prior to his first admission to hospital. Despite intensive medical treatment, both patients slowly deteriorated, and in both operation was performed in emergency sessions. In Mrs. N.D.G., resection started with the rectum and finished with the colon, whilst in Mr. H.F., an ileostomy was first constructed, and the colon was removed and finally the rectum.

#### SEE ILLUSTRATIONS ON PREVIOUS PAGE

FIG. V. Case 5: (i) the colon and rectum after excision. The disease involves the left half of the colon. There is considerable oedema of this portion of the bowel and extensive ulceration. The involvement lessens as the bowel is followed proximally; in the proximal half of the transverse colon, there are scattered ulcers, whilst, in the ascending colon and caecum, the mucosa has a normal appearance.

(ii) Close-up view of the rectum demonstrating the irregular ulceration of the mucosa and oedema of the bowel wall.

(iii) The transition in the mid-transverse colon; on the left side there is oedema of the colon wall and severe ulceration of the mucosa, whilst on the right side, the wall of the colon is thin, and only a few scattered ulcers are visible.

(iv) Close-up view of the caecum which shows no changes.



### RESULTS OF PRIMARY PROCTOCOLECTOMY IN THIS SERIES

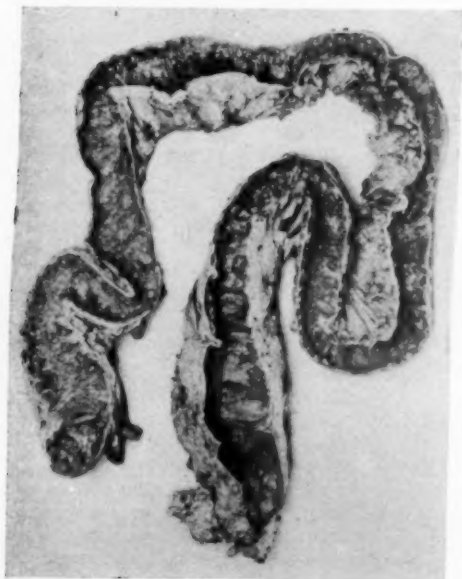
There was one death in this series; Mr. L.D. (Case 4) had progressed in a most satisfactory manner after the operation, but on the twelfth post-operative day he collapsed whilst having a bath and died a few minutes later. The cause was a massive pulmonary embolism.

In the remaining cases, the convalescence was largely uneventful; Mr. Mr.D. (Case 3) returned home on the 13th post-operative day, Mrs. J.R. (Case 1) and Mr. M.F. (Case 6) on the 21st post-operative day, and Mrs. A.L. (Case 2) and Mrs. N.D.G. (Case 5) on the 28th post-operative day. Apart from ileostomy stenosis, requiring minor adjustments, all have remained in very good health.

requirements operation can proceed. No special bowel treatment is needed because there is no soiling of tissues in this operation.

After induction of anaesthesia, the patient is postured on the operating table on the Lloyd-Davies' abdomino-perineal stirrups. The bladder is emptied, and a Winsbury-White self-retaining catheter inserted into the rectum, and retained there with a purse-string suture. The end of this tube is led away to a receptacle; this is a necessary step because manipulation of the colon and rectum during excision often forces liquid faeces through the anus, even though it has been occluded by a strongly tied suture.

In this series of cases a long, left paramedian incision was used. The ileostomy stoma was fashioned through a small trans-



(i)

FIG. VI. Case 6: (i) The colon and rectum after excision. There was gross involvement of the whole of the large bowel.



(ii)

(ii) A close-up view of the caecum and ascending colon. The mucosa was very friable and necrotic, and there were few areas in which the mucosa appeared viable. The muscle layer and serosa were oedematous and responsible for considerable diminution in the normal elasticity of the bowel.

### TECHNIQUE OF PRIMARY ONE-STAGE PROCTOCOLECTOMY

When the decision to operate has been made, further delay is minimised; after attention to electrolyte, protein and blood

verse incision placed below and to the right of the umbilicus.

These patients may be very ill and the operation should be planned and should proceed in such a way that the surgeon can

terminate it at a moment's notice, although this has not proved necessary in any case treated by us:—

1. If the colitis is most severe on the left side of the large bowel, the rectum is first excised and the dissection carried upwards along the colon. The operation can be concluded at any time by performing a temporary colostomy (Fig. 7 [i]).
2. If the colitis is generalized, and as severe on the right side as it is on the left, the dissection begins by constructing the ileostomy and the colon is then removed from the caecum in a distal direction (Fig. 7 [ii]).

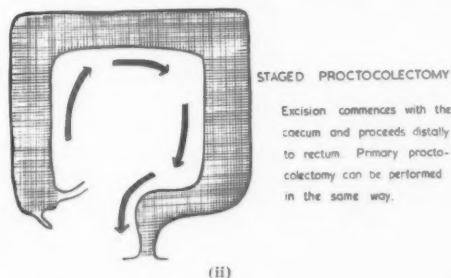
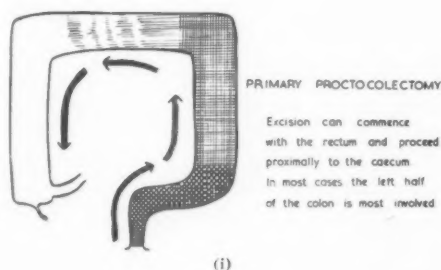


FIG. VII. (i) and (ii).

Maximum conservation of mesenteries and peritoneum has been observed, and, wherever possible, the raw surfaces of the peritoneal cavity have been re-peritonealized. The omentum has been preserved in each of these cases by dissecting it off the transverse colon. The space to the right of the ileostomy has been closed.

The rectum is excised in the usual way and the blood loss can be minimized by infiltrating the peri-anal and peri-rectal tissues with adrenalin in saline (strength, 1:250,000).

In each patient in this series, a skin graft was applied to the ileostomy at the conclusion of the operation. Our views concerning the skin-grafted stoma are given elsewhere.

#### DISCUSSION

It is impossible to save the colon in those patients with ulcerative colitis severe enough to require ileostomy. Attempts have been made to close the ileostomy, but Cattell (1953) reported that approximately half the

patients who had closure of the ileostomy had recurrence of symptoms necessitating a second operation within five years. Furthermore, Cattell (1953) followed a number of patients in whom ileostomy was performed and in whom it was thought that a subsequent ileostomy might at least be avoided, but persistent symptoms and the danger of malignant disease led him to abandon such a course. Counsell and Dukes (1952) report an incidence of 11.1 per cent. malignant development in 63 operated cases and in a later review Dukes (1954) observed cancer in 7 out of 120 operated specimens, an incidence of 5.8 per cent. Dukes (1954) summed up the position by stating "... a patient in whom

severe ulcerative colitis has lasted more than ten to fifteen years, should be regarded as having entered a period of life when the intestine becomes a definite risk." Few surgeons, therefore, believe that they are in a position to offer anything less than colectomy to a patient referred to him for surgical treatment of ulcerative colitis. One of the patients in this series, Mrs. A.L. (Case 2), had had a history of colitis extending back for twenty-five years, and operation revealed a carcinoma of the colon.

Opinion is not so well defined on the question of preservation of the rectum. A permanent ileostomy has been regarded as a serious disability, but it is not so bad as has been thought. Cattell (1953), Gabriel (1953), Naunton Morgan (1953) and Goligher (1953) have stated their views on the improbability of conserving the rectum, except in the exceptional case, and Goligher (1953) quotes several instances in which carcinoma developed in the rectum in association with ulcerative colitis. Brooke

(1953), restored bowel continuity in only one of sixty-four patients he subjected to surgery. Aylett (1953) was more optimistic, however, and suggested that if the colon is removed, the rectum is no longer bathed in mucinase, and so may recover because the protective layer of mucus is no longer destroyed. Brooke (1953), on the contrary, showed the fallacy of this reasoning by drawing attention to work which demonstrated that mucinase was also found in the small intestine. Aylett (1953) felt there was nothing to lose in attempting to preserve the rectum, because he argued that it could always be removed later. He stated that he had saved the rectum in nine cases, and in six of these resolution within the rectum occurred. Dunlop (1954) believes that in selected cases there is a place for ileo-rectal anastomosis. In our opinion, the rectum should be removed in ulcerative colitis; ulcers in the rectum heal by secondary intention with contracture of the lumen and the subsequent risk of malignancy is very real (Fig. VIII). We have performed ileo-rectal anastomosis when the involvement of the rectum is mild (one case), when the patient is very young (one case), and when, at colectomy for ulcerative colitis, a carcinoma is found with secondary deposits in the glands (one case).

The orthodox method for performing excision of the large bowel involves two or three stages. With chronic ulcerative colitis, ileostomy and colectomy are performed at the first stage and are followed by excision of the rectum, three to six months later (Cattell, 1953, 1955); in the fulminating case, an ileostomy is all that is done initially, and is followed by the colectomy and the excision of the rectum in two further stages. Recently, ileostomy and primary colectomy has been advocated for the acute fulminating cases and the procedure has gained substantial support. Crile and Thomas (1951) compare the colon in such cases with an area of extensive third degree burning and point to the frequency with which perforation is found at operation. Ripstein (1953) has found that in twenty-four hours 200-300 cubic centimetres of blood and 150-200 grams of protein may be lost from the ulcerated surface. Gardner and Miller (1951) reported seventeen cases of fulminating ulcerative colitis in which ileostomy and primary colectomy were performed; as soon

as the patient was well enough, the rectum was excised. Ripstein (1953) stated that he had treated forty-three patients with acute ulcerative colitis by a one-stage ileostomy, and colectomy to the recto-sigmoid junction, with but two deaths, one of whom was moribund at the time of the operation. Avery Jones (1953) and Goligher (1953) have both expressed themselves in favour of this procedure. In our opinion, an ileostomy can always be accompanied by a colectomy.

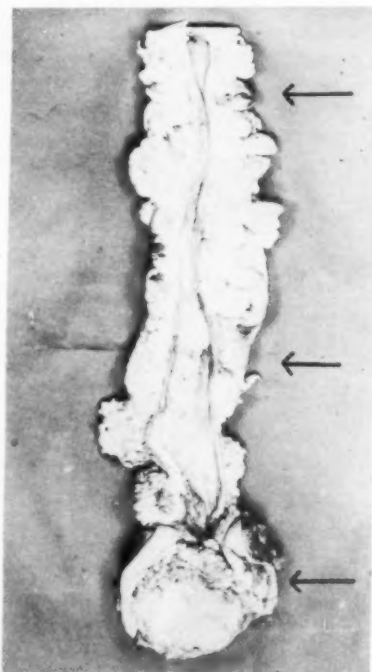


FIG. VIII. Rectum removed by abdomino-perineal excision eight years after ileostomy for ulcerative colitis. Three separate malignant tumours (adenocarcinoma) are indicated by the arrows.

In many cases of ulcerative colitis, the rectum shows the most advanced progression of the disease, and after ileostomy and colectomy, there still remains a considerable area of ulceration and infection. Even so, after colectomy, it is the rule for the patient to recover well, and after a month or two, to return to a good state of health in a way often not anticipated by the patient. Such patients are reluctant to return to hospital for excision of the rectum, and may postpone

further operation indefinitely. Excision of the rectum in these cases can prove a difficult dissection because of the deposition of excessive fatty tissue around a very contracted bowel, together with the partial or complete obliteration of tissue planes. For these reasons, a proctocolectomy offers theoretical and practical advantages over colectomy. Ravitch and Handelsman (1951) record a case so treated in their series. Goligher (1953) mentions five cases so treated at St. Mark's Hospital, and in a later paper (Goligher, 1954), nine more cases; all fourteen patients, treated by primary proctocolectomy, had smooth recoveries despite the inclusion of two gravely ill patients. In our own series, both acute and chronic cases have been treated by ileostomy and primary proctocolectomy. We have been impressed by the results and advocate the procedure in those cases in which the necessity for removal of the rectum is beyond question. It is an operation which requires careful pre-operative and operative planning, but for the surgeon familiar with excision of the rectum it presents no technical difficulties.

#### SUMMARY

1. Six cases of ulcerative colitis treated by ileostomy and primary proctocolectomy are reviewed:
  - (a) Two cases showed chronic ulcerative colitis, complicated by stricture formation. In one, the stricture was malignant.
  - (b) Two cases showed a sub-acute or chronic relapsing ulcerative colitis. In both, the history was long, and in both radiological studies at intervals showed proximal spread of the disease.
  - (c) Two cases showed an acute or fulminating ulcerative colitis. The history was short, and the patients gravely ill at the time of surgical intervention.
2. There was one unexpected death in the series: one patient died from a pulmonary embolism on the twelfth post-operative day. In the remaining cases, the convalescence was smooth.

3. In this procedure, the surgeon commences excision, either at the proximal end of the large bowel, or at the rectum, depending on the distribution of the ulcerative process. The dissection should proceed in such a way that the operation can be quickly terminated if necessary.
4. Primary proctocolectomy is advocated in those patients in whom an excision of the rectum will be required. Primary proctocolectomy achieves complete removal of the ulcerated area and at the same time avoids the necessity of the patient returning to hospital for what might seem to him an unnecessary operation, and for what might prove to the surgeon to be a difficult dissection.

#### REFERENCES

- AYLETT, S. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1032.
- BACON, H. E. and TRIMPI, H. D. (1950), *Surg. Gynec. Obstet.*, vol. 91, page 409.
- BROOKE, B. N. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1033.
- CATTELL, R. B. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1021.
- and COLCOCK, B. P. (1955), *Post Grad. Med.*, vol. 17, page 114.
- CORBETT, R. S. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1028.
- CRILE, G. and THOMAS, C. Y. (1951), *Gastroenterology*, vol. 19, page 58.
- DUKES, Cuthbert E. (1954), *Ann. Roy. Coll. Surg.*, vol. 14, page 389.
- DUNLOP, E. E. (1954), *Aust. N.Z.J. Surg.*, vol. 23, page 184.
- GABRIEL, W. B. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1035.
- GARDNER, C. and MILLER, G. G. (1951), *Canad. Med. Ass. J.*, vol. 63, page 370.
- GARLOCK, J. H. and LYONS, A. S. (1954), *Gastroenterology*, vol. 26, page 709.
- GOLIGHER, J. C. (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1025.
- (1954), *Ann. Roy. Coll. Surg.*, vol. 15, page 316.
- MILLER, C. G., GARDNER, C. McG. and RIPSTEIN, C. B. (1949), *Canad. Med. Ass. J.*, vol. 60, page 584.
- MORGAN, C. Naunton (1953), *Proc. Roy. Soc. Med.*, vol. 46, page 1036.
- RAVITCH, M. M. and HANDELSMAN, J. C. (1951), *Bull. Johns Hopkins Hosp.*, vol. 88, page 59.
- RIPSTEIN, C. B. (1953), *J. Amer. Med. Ass.*, vol. 152, page 1093.

## VAGINAL VAULT PROLAPSE

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**P**ROLAPSE of the vaginal vault may occur following hysterectomy, total or subtotal, performed by either the vaginal or the abdominal route. It is a distressing condition as far as the patient is concerned and often a source of anxiety and considerable worry to the surgeon called upon to restore the integrity of the supporting structures; indeed, it may tax his ingenuity to the utmost.

As a general rule, the prolapse makes its appearance soon after the original operation, therefore there is a tendency in some quarters to deride the surgical work which is followed by this unfortunate development. Inasmuch as the underlying principles of vault fixation following hysterectomy are now generally well recognized, it may be felt that this important step in the procedure has not received sufficient attention. This may be true in some cases but *prolapsus* may occur even in the most competent hands. In such circumstances there may be an inherent weakness in the supporting tissues which has not been clearly recognized.

Let us be quite clear at this juncture that abdominal hysterectomy *per se* has no place in the treatment of genital prolapse. To perform abdominal hysterectomy for this condition without attending to the stretched and torn vaginal muscles and fascia is worse than useless. No matter with what care the stump is fixed in the pelvic basin by attaching round ligament stumps, cardinal ligaments and the uterosacral ligaments, recurrence of the prolapse is a certainty. Subsequent procedures must include repair of the Pouch of Douglas hernia, which is often very large, in addition to plastic restoration of the vaginal fasciae and musculature.

It may be some little time after the abdominal hysterectomy before the symptoms of prolapse again show themselves, but there is nothing more sure than that they will do so eventually.

Vaginal hysterectomy is an operation which allows adequate pelvic repair to be performed in patients in whom there is some genital prolapse associated with a pathological condition requiring removal of the uterus. It is a procedure which requires clear anatomical visualization and adequate union of those supporting structures remaining in the pelvis after the uterus has been removed; furthermore, it demands some technical dexterity since the suturing is performed in a relatively confined space.

The technical details of a satisfactory operative procedure have already been described in a previous paper (Buchanan, 1953). Without undue repetition, it may be stated briefly that support is obtained by the formation of a "keel" of tissue, largely derived from the parametric pedicles extending from the round ligament stumps above to the uterosacral ligaments below. This "keel" is then interposed in the pelvic outlet as a firm support, particular care being exercised in the vicinity of the uterosacral ligaments.

In spite of meticulous care with fixation suturing, post-operative prolapse of the vaginal vault does occur at times, perhaps due to early absorption of ligature material or to low-grade post-operative sepsis. This second complication is apt to occur if the operator has been unwary enough to suture the keel of broad ligament and parametric tissues so tightly that necrotic sloughing occurs.

It is agreed of course that occasionally a patient presents with large uterine fibroids and an associated *prolapsus*. The tumours are too large for easy vaginal delivery and in such cases it is sound practice to perform primary vaginal repair and to follow this, either at the same sitting or later, with subtotal abdominal hysterectomy; care must be taken to secure adequate fixation of the cervical stump.



### *Clinical types*

There are certain well-defined clinical types of vault prolapse. Fortunately some of these are rather less difficult to control than others.

Type 1: Prolapse after subtotal hysterectomy, a comparatively normal cervix being present.

This usually presents as a gross cystocele and rectocele. In severe cases there is procidentia with the isolated cervical stump appearing outside the *introitus*. The button of cervix provides a minor fixation point for vaginal plastic repairs but will probably require further intra-abdominal suspension to secure an adequate result.

Type 2: Large enterocele (or Pouch of Douglas hernia) following abdominal hysterectomy with entirely adequate fixation of the cervical stump or the vaginal vault.

There is usually some relaxation of the vaginal outlet and the great depth of the Pouch of Douglas may have been overlooked at the time of operation. The enterocele presents on the posterior vaginal wall behind a well-defined cervix or vaginal vault and tends to become a progressive lesion.

Careful dissection in the posterior vaginal wall usually allows adequate demonstration of the hernial sac, which is stripped from the rectum posteriorly and closed at the neck behind the cervix by purse-string sutures which are so placed as to fix the new floor of the Pouch of Douglas to the back of the cervix (already well supported). The remainder of the sac is then excised and adequate posterior colpo-perineorrhaphy completes the procedure.

Type 3: Enterocele following vaginal hysterectomy.

This condition has been encountered where the anterior portion of the "keel" beneath the bladder is quite firm but an enterocele has developed, possibly due to some technical fault allowing separation of the uterosacral ligaments post-operatively. It could be due to the enterocele being overlooked at the time of vaginal hysterectomy—this would be most unlikely if the operator were at all experienced.

Such a case has been repaired by an approach through the posterior vaginal wall. The enterocele was easily opened and search made for the stumps of the uterosacral ligaments. Clinically recognizable ligamentous tissue was found and after excision of the redundant sac, these tissues were united firmly to the remains of the "keel." The vault after colpo-perineorrhaphy was firmly anchored.

Type 4: Vaginal inversion following total hysterectomy, abdominal or vaginal.

This is a difficult problem. The use of the Le Fort operation to occlude the vagina is unsatisfactory except perhaps in the aged; in any case it is not always successful.

Cases of this type require some form of abdominal fixation combined with further vaginal plastic procedures as indicated. Vaginal plastic repair operations depend upon restoration suturing, centred upon a well-supported vaginal vault. Hence the need in the standard Fothergill-Donald repair operation for accurate coaptation of the parametric tissues advanced in front of the uterine body, or in vaginal hysterectomy for close attention to the fixation suturing of the posterior portion of the "keel."

The principle that the prolapsed vaginal vault should be supported or suspended by artificial ligaments introduced abdominally is not a new one and has been practised with varying success and by various methods. The use of strips of ox fascia lata was described by Grant Ward in 1938.

The operative procedure to be described was developed after noting the suggestion by Shaw (1948) that strips of the rectus abdominis sheath be used as supporting structures for the inverted vaginal vault. The principles appeared to have a sound anatomical basis but the practice appeared to be somewhat tedious and difficult. Some alteration in technical approach was desirable and attempts were made to streamline the method.

Since 1948, 11 cases of vaginal inversion have been treated in this way. Reliance must not be placed entirely upon the vault sling. Vaginal plastic repair must be combined with it, either at the same time or as a two-stage operation. During the vaginal

work, particular care must be taken to eliminate the enterocele by opening and, when performing the posterior repair, obliterating the Pouch of Douglas.

Parsons and Ulfelder (1953), in their new publication, describe an operation of this type which however differs in some technical details from the procedure now to be described, particularly in the position of the slings in relation to the rectus abdominis muscle and in the method of introduction of the slings.

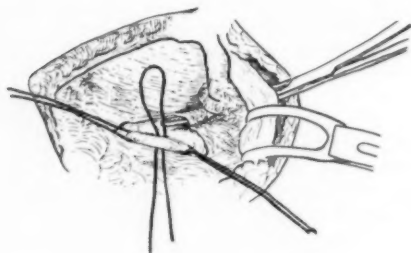


FIG. I. Diagram of appearance at operation showing the inverted vagina which has been elevated by two traction sutures and the peritoneal cuff reflected all round the vault. The mattress-type suture of waxed silk has been introduced for the right side and is being caught by curved forceps introduced extraperitoneally outside the rectus muscle (as in routine Gilliam-Crossen operation).

### Operation

1. After anaesthesia has been induced, the bladder is emptied by catheter. The inverted vagina is replaced manually and held in position by a multilayer gauze packing strip. An appreciable amount of this strip remains outside the vagina to allow easy extraction at a later stage.

2. The patient is placed in high Trendelenburg position and the abdomen is opened by midline subumbilical incision, the previous scar, if present, being cleanly excised. Devine's self-retaining retractor is introduced and the pelvis examined. Multiple fine adhesions may require attention.

3. The vaginal vault, being now clearly demonstrated by the packing, is caught at the two lateral angles in Morrison's forceps; the peritoneal covering is incised in a circular fashion and cautiously stripped back on all sides. There may be appreciable difficulty in

securing the bladder separation from the vaginal vault, and considerable care is necessary.

The lateral angles of the denuded vaginal vault are now caught in two stay sutures of black silk — the Morrison forceps being removed (Fig. I). The vaginal gauze pack is now removed by an Assistant.

4. The Devine's retractor is now removed and the anterior sheath of the rectus abdominis muscle separated from the underlying muscle about an inch and a half above the pubic symphysis as in commencing the Crossen-Gilliam suspension.



FIG. II. Photograph of an operation having appearances corresponding to those shown diagrammatically in Fig. I.

With light finger retraction and under vision a closed curved Kelly forceps is passed between the sheath and the rectus muscle to enter the plane of the transversalis fascia and progress downwards and outwards to the remnants of the round ligament. The instrument must now change its plane and be passed from this point inwards through the remnants of the broad ligament to emerge in the deperitonealized area on the vault. This is a tedious procedure at times and is sometimes materially aided by changing the Kelly forceps after the original tunnel is made for a Moynihan gall-bladder clamp with its quicker curve (Fig. II).

5. The clamp having been introduced extraperitoneally in this manner, now is made to grasp the loop of a waxed silk thread which has been introduced into the vaginal vault mattress-fashion and withdraws the loop

along the new extraperitoneal tunnel to emerge in front of the rectus abdominis muscle behind the rectus sheath.

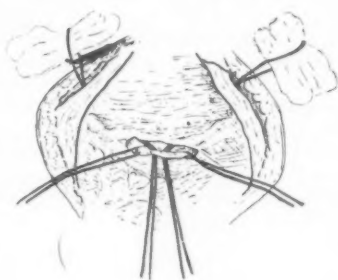


FIG. III. The loops of waxed silk have been introduced through the vaginal vault and withdrawn extraperitoneally on each side, appearing between rectus sheath and rectus muscle. These are now held preparatory to elevating fascial "sling" from the medial border of rectus sheath.

The same procedure is then carried out on the opposite side. Fig. III shows the waxed silk loops temporarily supported on gauze swabs.



FIG. IV. The fascial strips have been elevated from medial border of anterior rectus sheath and are now held preparatory to attaching loops of waxed silk for withdrawal along an extraperitoneal tunnel to appear in deperitonealized area.

6. Fascial strips are now to be raised from the anterior rectus sheath. The skin and fat of the abdominal wall are dissected back to expose the anterior sheath clearly. Parallel incisions are made one-third to one-half an inch from the main midline incision: the strips are raised from the underlying muscle by making transverse incisions just below the umbilicus and freeing the fascia from the muscle by clean dissection, down to the point of emergence of the waxed silk loops. The usual length of these strips is about four inches (Fig. IV).

7. The waxed silk loop on each side is now manipulated into a noose at its end: this is slipped over the free end of the appropriate fascial strip and tightened so that a firm grip is obtained. Gentle traction upon the ends of the waxed silk eases the fascial strip down extraperitoneally through its new tunnel so that it appears in the deperitonealized area over the vault. Similarly with its fellow of the opposite side.

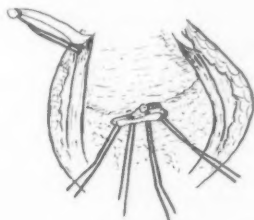


FIG. V. The right sling has been satisfactorily withdrawn along the extraperitoneal tunnel and appears in the vault area where it is now to be attached by thread sutures. On the left side the sling is caught in the waxed thread loop preparatory to withdrawal to the vaginal stump area. Fixation and closure of the peritoneal flap completes the procedure. The abdominal wall is closed with No. 32 stainless steel wire.

The attached ends remain firm in the rectus sheath and allow each strip to be sutured to the vaginal apex by interrupted silk sutures with a very nice appreciation of the tension required for adequate support. The reflected peritoneal covering of the vaginal vault is then repaired with plain catgut with particular care in the vicinity of the bladder.

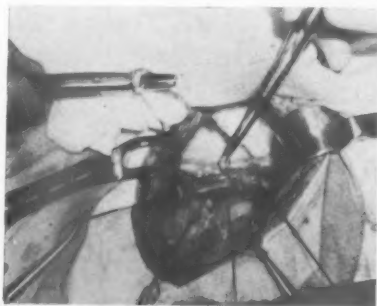


FIG. VI. Photograph of operative appearances corresponding to those shown diagrammatically in Fig. V.

8. The abdomen is closed in layers—suture of the remains of the anterior rectus sheath being effected with No. 32 stainless

steel wire. There is at times a good deal of tension in this plane and supporting tension sutures of silk-worm gut are often desirable.

9. Vaginal plastic repair should be practised either at the same sitting or at a later date. The usual repair based on sound principles is satisfactory but particular care must be taken to search for and remove any redundancy in the Pouch of Douglas.

This usually implies opening of the peritoneum in the posterior dissection and repair of the incipient herniation present. However, now that the vault is firmly anchored, this does not present any insuperable difficulty.

#### SUMMARY

A modified procedure for dealing with vaginal vault prolapse is presented as being

sound in principle. It produces an excellent cosmetic result and this method has been used with complete satisfaction in the treatment of 11 cases of vaginal inversion.

As the management of this type of case is always difficult, the employment of an operation of the nature just described is warranted by the prospects of permanent cure.

#### REFERENCES

- BUCHANAN, J. M. (1953), *Alfred Hosp. Clin. Rep.*, vol. 3, page 30.
- PARSONS, L. and ULFELDER, H. (1953), "An Atlas of Pelvic Operations." Philadelphia, W. B. Saunders Company, page 158.
- SHAW, H. N. (1948), *West. J. Surg.*, vol. 56, page 127.
- WARD, GRANT E. (1938), *Arch. Surg.*, vol. 36, page 163.

## DETERMINATION OF SHARPNESS OF CUTTING EDGES BY MICROSCOPIC EXAMINATION

By P. W. GOAD\*

**D**ETERMINATION of sharpness of cutting edges by microscopic examination is not novel in principle. Foster (1949) referred to photomicros of cataract knife edges taken in 1911, and the method has long been used by safety razor blade manufacturers in production control. Hillier (1951) used the technique of microscopic examination developed by Richards (1950) when preparing microtome knives for ultra-thin sectioning. Foster advocated the method for use in the field of ophthalmic surgery, in favour of the practical test much used by surgeons, that of trying instruments on stretched kid leather. He had found that cutting edges which were acceptable when examined microscopically invariably passed the kid test, but that the reverse did not always hold; mechanical imperfections and corrosion damage which did not affect the kid test were readily apparent under the microscope.

A number of more specialized techniques are available for investigating cutting edges. Kayser (1946) has mentioned the use of specially constructed optical goniometers and a diamond wedge interference method in the measurement of the angles of incidence of the minute facets that make up the extreme edge, and of the radius of curvature of the edge itself. Suitably adapted profilometers are capable of detecting scratches and imperfections 0.000001" deep on surfaces less than 0.001" wide. Honda in 1931 published an account of a test which involved moving the edge over strips of paper in a sawing motion, and Iwase (1954) developed a sharpness tester for razor blades which used a variable water level to bring the edge against a cotton thread, the force required to sever the thread being taken as a measure of the sharpness.

Probably the finest cutting edges produced with any degree of uniformity are found on safety razor blades. Surgical cutting instruments occasionally reach their high standard,

but are usually more difficult to manufacture because of their shape, and are often not required in sufficient numbers to call for the precision machines necessary for maintaining high quality.

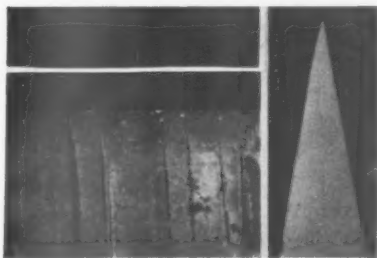


FIG. 1. High quality cutting edge on new detachable scalpel blade. (x 250)

Top left: Edge-on.

Lower left: Profile.

Right: Section at right angles to cutting edge.

According to Kayser and Foster, the sharpest cutting edges are made up of two mirror-like facets subtending an angle of 20-24 degrees, and having a radius of curvature at the extreme edge of approximately 0.00001". Good edges are never saw-like but are essentially straight, even when examined at a magnification of x 500. The final facets (the facets adjacent to the cutting edge) are for all practical purposes free from scratches, and the few present are less than 0.00001" deep.

The foregoing is sufficient to define a high quality cutting edge and it forms a useful basis for a sharpness standard.

### APPLICATION OF METHOD

The microscopic method of examination for sharpness was applied by the author to detachable scalpel blades and safety razor blades in the course of a corrosion investigation. It was found that an accurate estimate of sharpness could be obtained by examination of a cutting edge in two positions, profile and edge-on, at a magnification of x 500. No

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precise measurement could be made of the radius of curvature of the extreme cutting edge, or the depth of scratches in the final facet, but an optical comparison of these features could readily be made with those of cutting edges of known high quality. For examination of the contours of the edge facets, a polished microsection was prepared in a plane at right angles to the cutting edge. The microscope used was an inverted-stage metallurgical type, and all examinations were carried out with vertical illumination.

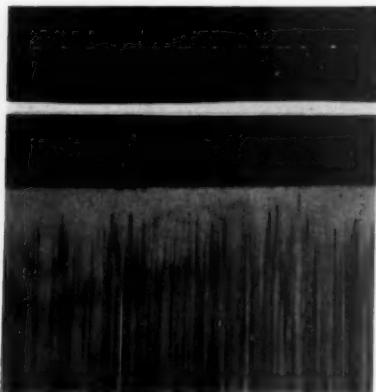


FIG. II. High quality cutting edge on new safety razor blade. (x 360)  
Top: Edge-on.  
Lower: Profile.

Kayser and Foster have objected to the use of vertical illumination when making profile examinations on the grounds that the sloping facets reflect the light away and give a misleading appearance to the edge. This objection was overcome by supporting the blade on a small piece of plasticene and manipulating it until the final facet was at right angles to the optical axis of the microscope. This was simply carried out by observing the edge through the microscope and noting the necessary movement of the objective to bring any part of the field into focus, and altering the setting of the blade accordingly.

For examination in the edge-on position, the blade was supported by pushing one end into a piece of plasticene. It was not found necessary to have the blade in perfect alignment with the optical system; slight deviations

had little effect on the appearance of the edge. Grease and dust were removed from the blade by wiping gently with cotton wool damped with carbon tetrachloride.

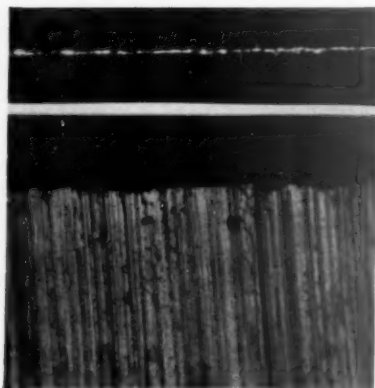


FIG. III. Poor quality cutting edge on new detachable scalpel blade, showing saw-tooth effect produced by coarse grinding of final facet. (x 360)  
Top: Edge-on.  
Lower: Profile.

For examination of an edge in section, the blade was clamped between steel strips and polished by the usual metallographical techniques.

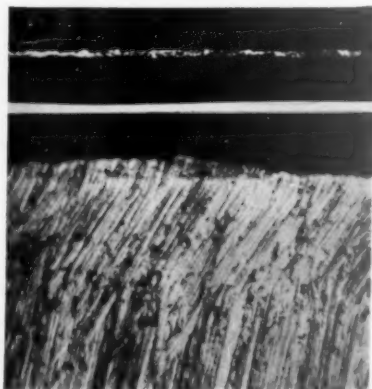


FIG. IV. Poor quality cutting edge on used, and hand re-sharpened detachable scalpel blade, showing "feather edge," half broken off (out of focus, bending backwards). (x 360)  
Top: Edge-on.  
Lower: Profile.

## RESULTS OF EXAMINATION OF TYPICAL CUTTING EDGES

The microscopical appearance of two high quality cutting edges, one on a detachable scalpel blade, the other on a safety razor blade, is shown in Figs. I and II. In the profile views, the final facets are shown to be smooth and scratch-free and the cutting edges straight. The edge-on view shows a thin uniform line. The sectional view, in Fig. I, shows the final facets converging at an angle of approximately 23 degrees.

The edge-on and profile appearances of a blade of poor quality are shown in Fig. III. The coarse grinding operation which completed the sharpening operation of the blade produced a saw-toothed effect at the cutting edge.

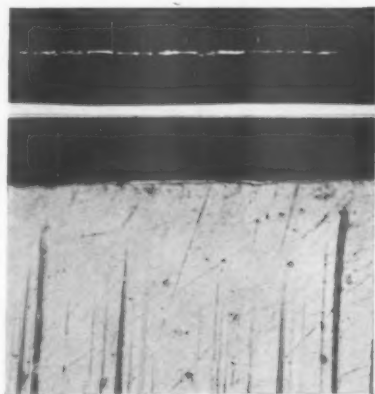


FIG. V. High quality cutting edge on detachable scalpel blade, after being used once, showing blunting by turning over of the extreme edge. (x 360)

Top: Edge-on. Lower: Profile.

Hand finishing of edges often results in the formation of an unstable "feather edge," which is liable to break off or bend over with the slightest use. A typical feather edge on a scalpel blade which had been hand re-sharpened by hospital technicians is shown in Fig. IV. Feather edges are readily visible at low magnifications, x30 or less; they stand out, as their name implies as a kind of fringe on the edge proper. Stropping on leather or on the palm of the hand will partly remove a feather edge, but usually a poor quality cutting edge remains.

Blunting of a good quality steel blade occurs by nicking or turning over of the

cutting edge due to accidental contact with hard objects rather than by actual wearing away of metal. A scalpel blade blunted during normal hospital use is shown in Fig. V.

## CONCLUSION

The microscopic method of examination for sharpness is simple, does not call for special equipment, and is non-destructive except when a microsection is occasionally prepared. The quality of the entire length of the cutting edge can be rapidly assessed and experimental variables are few.

Inverted stage metallurgical microscopes with built-in vertical illumination systems appear to be the most suitable instruments for making the examinations, but hand microscopes can be adapted in the manner described by Foster, or by Richards.

Durability of a properly prepared edge depends on the quality of the blade material and its heat treatment. These usually have to be taken for granted, but examination of a sample of a batch or brand for microstructure and hardness would give a good indication of ability to maintain sharpness in service.

## SUMMARY

A method of determination of sharpness of cutting edges by means of microscopic examination at x 500 is described. Accompanying illustrations show the microscopic appearance of cutting edges of various qualities. A definition of sharpness and other methods of determining it are outlined.

## REFERENCES

- FOSTER, J. (1949), *Trans. Inst. Brit. Surg. Technicians*, vol. 6, page 47.
- HILLIER, J. (1951), *Rev. Sci. Instruments*, vol. 22, page 185.
- IWASE, K. (1954), *Metal Progress*, vol. 65, page 112.
- KAYSER, J. F. and FOSTER, J. (1945), *Proc. Roy. Soc. Med.*, vol. 39, page 835.
- RICHARDS, O. W. (1950), *Rev. Sci. Instruments*, vol. 21, page 670.

(The above study was begun as the result of enquiries made by several surgeons into the possibility of obtaining sharper knives. A necessary step was the production of a method to demonstrate degrees of sharpness.

The paper is included in this Journal because the necessity for a scientific approach by way of a particularly valuable adjunct to surgical technique is becoming recognized.—Ed.)

## PARAFFINOMA OF THE BREAST

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THE lengths to which women will go to enhance their appearance with the object of attracting the attention of the opposite sex, are sufficiently well known as to occasion no surprise. In the cases described here, an artifice is invoked which adds yet another entity to the list of more frequently encountered lumps in the breast. Paraffinoma of the breast is an apt descriptive term for the condition which results from the injection of either low melting point paraffin wax, liquid paraffin and beeswax or a mixture thereof into the mammae for the cosmetic purpose of increasing the bust line.

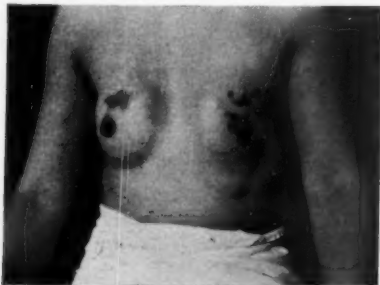


FIG. I. Patient with bilateral paraffinomata of breast displaying the multiple pigmented scars of temporary healed sinuses.

During the past 3 years seven examples have come to our notice. It is presumed that the practice of injecting the breast with paraffin is more widespread than this small series of cases would appear to indicate, as the patients presented themselves only when untoward complications of the procedure forced them to seek medical advice.

The pathogenesis of the condition in each patient is rather similar, and may be typified and best described by quoting the case history of one at length.

### CASE HISTORIES

#### Case 1

Mrs. H. is a married Chinese woman aged 41 with two children. During the last war when her husband was out of work she became employed as a cabaret

girl in Macao. Six years ago, a nurse of old acquaintance suggested to her that she have the contour of her bust enhanced by a series of injections which she was prepared to carry out. The nurse, apparently, had had the procedure performed on herself and recommended it to the patient as a means of improving her business as a taxi dancer.

At one session each breast received six injections placed circumferentially. Paraffin wax heated beforehand was used for the injections which were understandably painful. Some bleeding occurred from the puncture sites. During the following week the breasts continued to be painful. The desired enlargement of her breast was however achieved and she was well pleased with the result. The breasts felt heavy but there were no symptoms experienced during menstruation. For six months her pleasure in the cosmetic effect remained unsullied until multiple small vesicles developed in the skin of the breasts and discharged white material which the patient likened to coconut scrapings. These sinuses alternately healed and broke down. Fresh ones appeared from time to time and eventually the colour of the discharge changed to yellow. For a long time she tolerated the inconvenience and discomfort of the discharging sinuses as a sense of shame disinclined her to seek treatment, but she was finally persuaded to have the breasts attended to.



FIG. II. Post-operative result of bilateral simple mastectomy and immediate split skin grafting for paraffinomata.

On examination the patient was seen to be an otherwise healthy woman. Over each breast the skin was studded with small puckered pigmented scars of sinuses in a temporary healed phase (Fig. I). There was no nipple retraction or discharge and the breasts were regular and symmetrical in contour. Each breast was occupied by a nodular mass, which occupied the whole extent of the organ with the exception of the axillary tail, and of a vulcanite rubber consistency. Firmly attached to the overlying skin the masses were movable on the deeper

structures and the breasts were not tender to palpation. There was no associated axillary gland enlargement.

A bilateral simple mastectomy was carried out in separate stages and included removal of the nipples and the scarred mammary skin leaving large raw areas of chest wall for immediate split skin grafting (Fig. II).

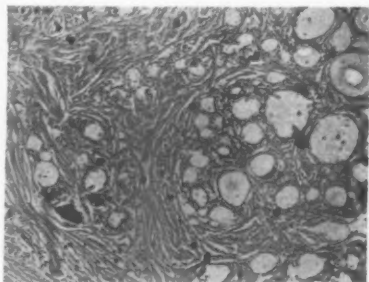


FIG. III. Histological appearance of mammary paraffinoma showing replacement of breast tissue by fibrosis, round cell infiltration, giant cells and cystic spaces which contained the injected material.

On cut section the operation specimens presented a greyish white homogeneous appearance and imparted a sensation like that of crude soap to the hand. Microscopic examination revealed virtually complete replacement of the normal breast histology with fibrous tissue and aggregates of cystic spaces which contained deposits of paraffin wax. Giant cells of the foreign body type were interspersed in groups amidst a general background of small round cells, the whole histological picture being one of a chronic granuloma to which the term paraffinoma seems appropriately applicable (Fig. III).



FIG. IV. Migrant paraffinoma tissue outlined by skin marking.

In two patients, migration of the wax within the layer of superficial fascia from the breasts on to the anterior abdominal wall was a feature of interest. This phenomenon is illustrated by the experience of one patient whose case history is as follows.

### Case 2

Three years previously Mrs. Y., then aged 40, had both breasts injected with paraffin for cosmetic reasons. For a time following the injections some oily fluid leaked out from the puncture sites but eventually ceased spontaneously. Two years later she noted the appearance of nodular swellings on the abdominal wall and three months after gradual increase in the size of both *labia majora* but mainly the right. At the same time as the labia enlarged, reciprocal decrease in the size of the breasts was noted. The whole process was painless.

The nature of the nodular masses was confirmed histologically as being paraffinoma tissue by biopsy, and their distribution in the abdominal wall and labia are illustrated by skin marking in Fig. IV.



FIG. V. The post-operative result of bilateral simple mastectomy and fasciectomy of the anterior abdominal wall in one patient in whom migration of paraffinoma had occurred extensively.

No treatment was advised for this patient as she was not experiencing any symptoms. Operative treatment was, however, carried out for another woman in whom widespread migration of wax had occurred, because she was so very worried about her condition. Bilateral simple mastectomy and extensive subcutaneous fasciectomy of the abdominal wall in stages was required to remove all the paraffinoma tissue (Fig. V).

One patient died as a result of having her breasts injected with paraffin, the complication being fat embolism in this case.

### Case 3

Two days previously Mrs. T., aged 32, had been given paraffin injections into the right breast. One hour later she experienced headache and giddiness

and lapsed into unconsciousness four hours after the injection, becoming incontinent of faeces and urine. Initially treated in a private clinic she was transferred to hospital where she was found to be semi-conscious and restless. Three puncture marks were seen on the right breast. Her pupils were small, equal and reacted to light sluggishly. Muscle tone was normal but her tendon jerks were brisk and the Babinski response positive bilaterally. Respirations were shallow and rapid and physical signs of basal pneumonia detected. Her pulse was rapid and weak with a B.P. 95/60. The patient became progressively more comatose and died the day after admission to hospital. A clinical diagnosis of fat embolism was confirmed at post-mortem examination.

### DISCUSSION

That this attempt to achieve a Venusian ideal is fraught with untoward consequences is emphasized by the above description of sinus formation, migration of paraffin and oil embolism. However it is not so much remarkable that complications occur as that the breast tissue tolerates at all, for any length of time, the injection of a liquid foreign body into its substance. Certainly breast paraffinoma have been an incidental finding in a few patients undergoing treatment for other conditions in whom the injections have apparently been quite innocuous.

What determines the success or otherwise of the procedure is most probably, if not wholly, related to the original injection technique. Clearly the intravascular injection of liquid paraffin producing oil embolism is a hazard easily avoided by an operator aware of the possibility. We can only suggest the mechanism of sinus formation and migration. The former occurs if any of the injection is placed too superficially. Paraffin deposited between the breast parenchyma and skin provokes a fibrous tissue reaction which tethers the skin to the underlying gland at the injection sites forming a track along which the paraffin can subsequently surface. Low grade chronic infection is also another likely factor governing the appearance of sinuses as these injections are invariably carried out by persons of doubtful education in the principles of asepsis.

In the instances of migration, gravity undoubtedly influences the spread, since movement occurs downwards on to the abdominal wall. The *modus operandi* is explicable on anatomical grounds. The breast has no true capsule; its lobules are situated within the superficial fascia of the chest wall. This

fascia, as it continues over the anterior abdominal wall differentiates into two layers, a superficial fat-containing layer and a deeper fibrous layer. At the inguinal region these fascial planes of Camper and Scarpa fuse with the deep fascia and with the skin in a linear attachment which gives rise to the so-called flexor crease of the hip joint (Wood Jones, 1919). It thus transpires that paraffin, unconfined by a gland capsule can escape from the interstices of the breast and gravitate unhindered, like rain water percolating through top soil, within the superficial fascia until further progress is checked in the groin, making the external genitalia the lowest point to which descent can occur. The process is gradual, taking several months, and during its movement the paraffin stimulates a chronic inflammatory response at descending levels leaving behind it a fibrous tissue reaction to mark its progress.

A point of interesting speculation concerns the possibility of malignant change appearing in breasts, the seat of paraffinoma.

Hydrocarbon compounds have acknowledged carcinogenic properties, for example, in shale oil workers cancer and benzanthracene-induced cancer in mice; and in paraffinoma of the breast, a potentially carcinogenic agent is wilfully brought into intimate contact with an organ of high spontaneous cancer incidence thus creating circumstances of sinister significance. Nevertheless although carcinoma of the breast is common among Chinese women, we have seen no case in which it could be related to paraffin injections, or in which paraffinoma were also present.

### SUMMARY

1. Examples of paraffinoma of the breast following intramammary injection of liquid paraffin are described.
2. Case histories are recorded of patients exhibiting complications of the procedure which include sinus formation, migration of paraffin and oil embolism.
3. The pathogenesis of these complications is discussed.

### REFERENCE

- WOOD JONES, F. (1919), "Principles of Anatomy as Seen in the Hand." London, Bailliere Tindall & Cox.



## TIDAL DRAINAGE OF THE BLADDER

By I. A. FERGUSON

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SINCE Munro (1935) first described a method of tidal irrigation of the bladder, numerous modifications have been developed. These modifications have been mainly efforts to make the apparatus more simple and, with few exceptions, have not altered the main principle involved.

### THEORY OF TIDAL DRAINAGE

One way to visualize the manner in which a simple apparatus works, is to imagine it being built up in stages, beginning with a tube leading straight from a reservoir and drip chamber to the bladder, as in Fig. I. Fluid passing in would fill the bladder until the pressure in it equalled the head of pressure in the reservoir.

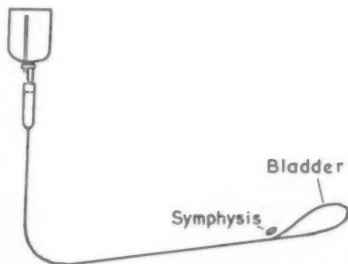


FIG. I. Illustration of direct connection between flask to bladder (with rubber tubing).

Now add a syphon to the tube as in Fig. II and begin to fill the bladder. As the bladder fills, the pressure within it will show as a head in the up-limb of the syphon. It will continue to fill until the bladder pressure is equivalent to the height of the crest of the syphon, at which point fluid will run over and flow out of the system. Provided the rate of flow from the reservoir and kidneys is not greater than the capacity of the syphon, this syphon will now empty the bladder. However, once the bladder is empty it will remain empty, as there is no means of breaking the syphon.

Now add an air inlet to the system at a point higher than the crest of the syphon (so that fluid will not leak out). The easiest way of doing this is to add a vertical tube, with its open end higher than the crest of the syphon, as in Fig. III. When the bladder begins to fill, this time, the bladder pressure will show as equal heads in the up-limb of the syphon and in the air inlet tube. Filling and emptying will now occur as in the last system but this time it would appear that, after emptying is complete, air will be drawn into the syphon via the air inlet; the syphon will break, and the system will be ready to begin a new cycle. Though there are several ways in which the air inlet, syphon, and the remainder of the tubing can be arranged, this apparatus we have built up is the basic tidal drainage machine.

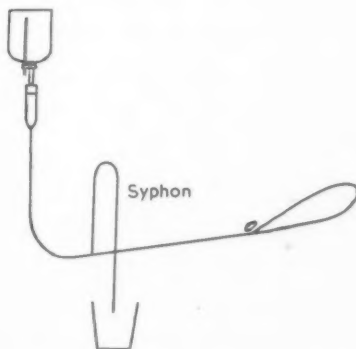


FIG. II. Syphon added to system illustrated in Fig. I.

The apparatus illustrated in Fig. III would work well if there were no such things as resistance to flow or surface tension. It was soon found that such a machine would always fail to empty the bladder owing to premature breaking of the syphon. An attempt was made to overcome this by slowing the rate at which air could enter the system. This was

done by making the actual point of air entry a fine capillary tube or, as in Riches' (1943) apparatus, by clipping the rubber tube joining the glass air inlet tube to the main line of tubing. This method would work but was, in fact, for that purpose an unnecessary complication.

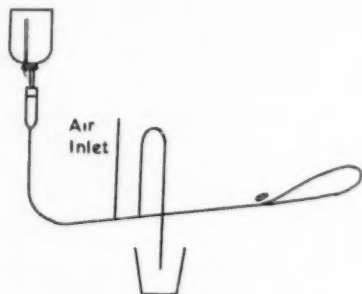


FIG. III. An air inlet has been added to the system of Fig. II. The diagram is now an illustration of the basic tidal drainage apparatus.

The other difficulty encountered in this basic apparatus was that of air-locking in the syphon tube—an effect of surface tension. It was found that instead of breaking the syphon cleanly, air invariably entered in bubbles and passed into the syphon as alternating columns of fluid and air, thus breaking the syphoning action while a considerable quantity of fluid was still in the tubing. As the machine fills at the beginning of the next cycle, this fluid is displaced into the down-limb of the syphon, and its weight lifts the ascending column in the up-limb by a corresponding amount, producing discharge of the fluid in the apparatus at a pressure correspondingly lower than that at which it was set. Discharge in these machines thus can, and often does, occur at a pressure as much as 10 to 15 cm. below the set pressure. Where the syphon cannot be set higher than 10 cm. above the pubic symphysis, such a machine will often fail to put any fluid at all into an atonic bladder.

After giving some thought to the subject of hydrodynamics it was realized that there was a pressure gradient along the tube from bladder to discharge point and that, as a result of this, there was a critical level below that of the pubic symphysis such that, if

the air inlet into the syphon was placed below it, the bladder had to be empty before the syphon could break. At the same time it was realized that a U in the air inlet would lower the effective air inlet pressure without the necessity of lowering the actual point of entry into the main tubing. The reasoning behind this is set out clearly in the paper of Miller, Payne and Lance (1954).

The next problem to solve was that of air locking. This was found to be more difficult and progress came to a halt for some time. Finally, it was noted that some machines would at times discharge without air locking. The reason was searched for and was found to lie in the resilience of the tubing used. When this was discovered it was only a short step to add a rubber diaphragm to the system to increase this resilience. This was found, after a little experiment, to eliminate air locks effectively. This and the previous modification of the basic machine are shown added in Fig. IV. This diaphragm is the major new modification described in this paper.

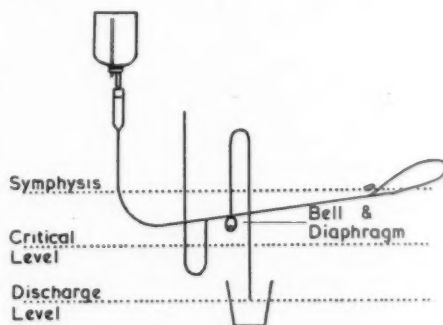


FIG. IV. The basic apparatus has now been altered to include modifications. These are:

- A rubber diaphragm has now been added to the system.
- A loop has been placed in the air inlet so that the pressure in the system has to fall below the critical level before air can enter; this ensures emptying of the bladder.

#### A PRACTICAL FORM OF THE APPARATUS

A connection was designed that joined the two T-tubes and the diaphragm bell into one unbreakable fitting (a most important consideration in any apparatus meant to be used in the wards of a hospital). The fitting, with its dimensions, is illustrated in Fig. V. It was made from brass tubing of appropriate



the emptying cycle is then complete. As the machine fills, the air trapped in the body of the connection between the air inlet point and the top of the connection is forced through the syphon by the rising fluid level, thus cleaning out any fluid remaining in the tubing; the cycle begins again.

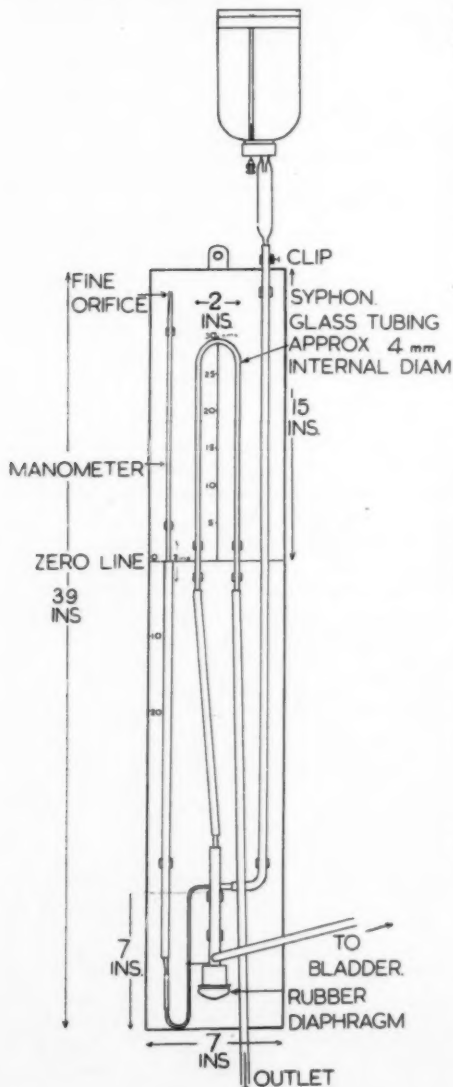


FIG. VI. Scale drawing of the arrangement of the apparatus as a whole on a back-board.

The apparatus in the form described has been in use for over a year, and has proved satisfactory.

#### *Points in the use of the apparatus*

The maintenance of the zero line of the machine in accurate line with the patient's symphysis is of great importance, especially in those patients who have a recent spinal cord injury and an atonic bladder. It must be realized that a pressure change of only 5 or 6 cm. of water may make the difference between an empty and an adequately filled bladder, and equally minor pressure changes may overdistend it. A back toilet and making the patient's bed may raise or lower him up to 8 cm. or so, quite upsetting the filling of his bladder. It can be seen then that after each time the patient is moved the machine must be releveled with some form of straight edge. In the later stages, as the bladder becomes more and more tonic and contractions begin to occur, this level becomes gradually less critical.

Another point that is important and easily overlooked is the relation between the end of the discharge tube and the surface of the fluid in the discharge bottle. Should the end of the discharge tube be allowed to fall below the surface of the fluid, then the discharge pressure will be raised by the amount it is below the surface, which could be as much as 15 or 20 cm.

In the machine herein described it is important to note that it cannot work if the end of the discharge tube is allowed to remain above the level of the air inlet U-tube. The syphon then cannot break.

Finally, though it may seem obvious, it is also important to realize that no machine can function properly with a blocked or partly blocked catheter.

#### SUMMARY

1. The principle of the tidal drainage machine is reviewed.
2. A further modification of the basic apparatus in the form of the addition of a rubber diaphragm is described.
3. Some difficulties in the operation of these machines are mentioned.

## ACKNOWLEDGEMENT

In conclusion I would like to express my gratitude to Mr. K. C. Bradley, without whose help and encouragement this article would not have been written.

## REFERENCES

- MILLER, W. M., PAYNE, J. T. and LANCE, R. V. (1954), *Surg. Gynec. Obstet.*, vol. 98, page 413.  
MUNRO, D. and HAHN, J. (1935), *New Engl. J. Med.*, vol. 212, page 229.  
RICHES, E. W. (1943), *Lancet*, vol. 2, page 128.

## CYSTIC CHANGES IN A LATERAL SEMILUNAR CARTILAGE IN A BOY OF 5 YEARS

By G. G. McCLOSKEY

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**C**YSTS of the semilunar cartilages of the knee are not common in children and are rare in the early years of life. A child of four years described by Infante and Santervas (1943), is the youngest case yet reported. Sjoval (1942), has reported a cystic cartilage in a child of five years and Ollerenshaw (1929) one in a six-year-old child.

Approximately 80% of cysts of the semilunar cartilages occur in the lateral meniscus, and occur on the external border of the mid portion of the cartilage. They are multilocular, are filled with a mucoid material and are usually multiple, varying from minute spaces to an inch or more.

A definite history of trauma is obtained in half the cases, the trauma usually being mild and the cyst tending to appear from one to six months after the trauma. The swelling rapidly increases to a certain size and then remains stationary. The cyst is rounded, oval tense and with the long axis horizontal in the joint line immediately over the meniscus. There are no signs of inflammation. The swelling becomes slightly less tense when the knee is flexed. The cyst is only slightly painful on pressure and usually gives pain only on extreme flexion or extension; the commonest complaint being of a dull ache and interference with the movement of the knee. The cysts are more common in males and occur chiefly in the second and third decade.

### CASE REPORT

L.A., born on 2nd November, 1948, first presented on 29th June, 1953. His mother complained that he had been limping with his right leg for the previous four weeks, and that the right leg would not straighten fully. There was no pain. No history of trauma was obtained.

On examination the child was seen to walk with a mild limp. The right knee lacked the last 10° of extension due to a painful block and the hamstrings were in spasm. Other movements were full and there was no swelling of the joint. There was no wasting of the quadriceps femoris. X-ray examination showed no abnormality. He was admitted to

hospital for investigation, which proved negative. Immobilization did not alter the clinical findings. Later it was noticed that a definite click was palpable on the lateral side of the right knee joint, over the joint line, as the knee moved through the position of mid-flexion. Accordingly he was readmitted for exploration of the joint.

At operation the lateral cartilage was seen to be malformed. It was broader than usual and had a small flap attached to the inner aspect of the anterior horn. The cartilage was removed and it was seen that there were several small cysts present on the outer border of the cartilage in the region of the middle 1/3rd. They were 1-2 mm. in diameter.

The pathological report on the sectioned cartilage reads:

"The tissue is fibro-cartilage and cysts of various sizes are present in its substance, the largest being 0.5 cm. in diameter. They contain a thin gelatinous material. Histological examination reveals numerous cysts, the majority of which have a clearly defined lining of flat cells.

"Apart from the presence of cysts, the cartilage has a normal appearance."



FIG. 1. Photomicrograph showing a cyst with a clearly defined lining of flat cells.

### DISCUSSION

Cysts of the semilunar cartilages were first described by Ebner in 1904 but it was not until 1921 when Ollerenshaw described a small series that they appeared in the English literature.

The German view was that these cysts were due to a process of degeneration occurring in the cartilage and caused by deficient nutrition



following trauma. Ollerenshaw (1921, 1929 and 1935) postulated the view that the cysts were embryological in origin, developing from pre-existing spaces in the cartilage and described the lining cells as being endothelial.

However, the congenital nature of these cysts is denied by most workers and the lining membrane has been shown to be synovial in nature and not endothelial. Many other theories have been postulated.

1. Trauma is followed by a reaction in the injured region, mucoid degeneration of the area occurs and the fabrication of a mesothelial cyst wall is brought about by compression of the surrounding fibro-cartilage (Taylor, 1935).
2. Obliterative changes in the artriotes of the paramemiscal tissue.
3. Trauma and congenital anomaly.
4. Synovial inclusions.
5. Synovial implants into cartilage at the time of trauma (Zadek and Jaffe, 1927).
6. Lymphatic in origin (Kleinberg, 1927).
7. Degeneration of cartilage (Allison and O'Connor, 1926).

King (1931, 1940) has shown that in certain areas of the body, as a result of an active stimulus, cells become enlarged and a mucinoid material accumulates at the same time. When the active stimulus discontinues, the process ceases and the cells at the periphery become, or those which have been but slightly affected remain, spindle in form and the

central material is thus surrounded by tissue which is of the synovial type. By studying the cells in the so-called degenerating areas King was able to show that these changes were due to cellular activity and not to degeneration.

We believe that in this case, a congenital malformed cartilage became subject to mild chronic trauma and that this trauma acted as a stimulus which caused the cells of the cartilage to develop, secrete and thus form cysts.

#### ACKNOWLEDGEMENT

I wish to thank Mr. J. B. Colquhoun, senior Orthopaedic Surgeon, Royal Children's Hospital, Melbourne, for permission to publish this case.

#### REFERENCES

- ALLISON, N. and O'CONNOR, D. S. (1926), *Surg. Gynec. Obstet.*, vol. 42, page 259.
- INFANTE, C. and SANTERVAS, C. (1943), *Rev. Soc. Paediat. Rosario*, vol. 8, page 99.
- KING, E. S. J. (1931), *Surg. Gynec. Obstet.*, vol. 53, page 606.
- (1940), *Surg. Gynec. Obstet.*, vol. 70, page 150.
- KLEINBERG, S. J. (1927), *J. Bone Jt. Surg.*, vol. 9, page 323.
- OLLERENSHAW, R. (1921), *Brit. J. Surg.*, vol. 8, page 409.
- (1929), *Brit. J. Surg.*, vol. 16, page 555.
- (1935), *Brit. J. Surg.*, vol. 23, page 277.
- SJOVALL, H. (1942), *Acta Chir. Scand.*, vol. 86, page 561.
- TAYLOR (1935), *J. Bone Jt. Surg.*, vol. 17, page 588.
- ZADEK, I. and JAFFE, H. L. (1927), *Arch. Surg.*, vol. 15, page 677.

## UNUSUAL PENETRATING PEPTIC ULCERS

By EDWARD WILSON

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THE all too familiar typical penetrating peptic ulcer involves the pancreas; but, it is with three ulcers which had involved not the pancreas, but, instead, the liver, spleen, and common bile duct, that this brief communication is concerned. Other viscera may, of course, be involved by a penetrating ulcer but such is extremely rare.



FIG. I. The excised ring of tissue consisting of the edges of the ulcer which was penetrating into the liver. The scale is in inches.

In Case 1 (a man of 37 years) the ulcer was two and three-quarter inches in diameter (Fig. I) and extended into the under surface of the liver. It had arisen from the lesser curvature of the stomach near the cardiac orifice, and although he had a history of a peptic ulcer for some 10 years the radiologist concluded from the position, size, and fixation of the ulcer that now it was probably malignant. This was not impossible for the pain was no longer affected by diet and antacids and he had lost two stone in weight in the past month. At operation, however, when the ulcer was examined directly across the lumen of the stomach it appeared benign. Since his general condition was poor and since considerable dissection was necessary to display the ulcer, operation was restricted to excising the edges

of the ulcer and to closing the opening in the stomach. A drain tube was inserted down to the region of the floor of the ulcer which was left *in situ*. Any gastric resection at that time would certainly have been fatal. Microscopic examination of the excised edges of the ulcer showed it to be a chronic gastric ulcer.



FIG. II. Chronic gastric ulcer penetrating deeply into the spleen.

In Case 2 (a woman of 47 years) increasing upper abdominal pain had been present for six months. This pain was not related to meals, but was partially relieved by antacids. Ten pounds weight had been lost during those six months. An opaque meal examination showed "a filling defect 3 inches in diameter with ulceration on the upper part of the greater curvature of the stomach," and it was reported that "this was almost certainly a carcinoma." At operation the spleen and stomach were found to be firmly adherent and were removed *en bloc* (Fig. II). There was no evidence of any metastasis. After a partial gastrectomy had been performed it was considered most likely that the lesion was a benign gastric ulcer penetrating the spleen and that the filling defect was due to hypertrophic mucosa around the ulcer. The actual ulcer crater was  $1\frac{1}{2}$  inches in diameter. Continuity of the alimentary tract was restored in the manner of the Billroth I procedure. The benignity of the ulcer was later confirmed by microscopic examination.

In Case 3 (a man of 45 years) a chronic peptic ulcer of the duodenum had penetrated into the common bile duct. This man was known to have had a peptic ulcer for at least 10 years; and he had always had relief of the epigastric pain with antacids and milk. However, when a mild attack of jaundice and pain beneath the right costal margin occurred he was referred for investigation. Cholecystography failed to show a gall-bladder shadow, but an opaque meal examination showed an ulcer of the posterior wall of

the first part of the duodenum. It is of note that no barium then passed into the biliary tree. On the other hand, by the time of the operation a month after the jaundice had increased and a definite choledochoduodenal fistula had developed. (This was not an artefact due to operative trauma!) Treatment consisted of separating the common bile duct from the duodenum, inserting a T-tube into the duct for three weeks, excising the duodenal ulcer, and closing the opening into the duodenum. The preliminary part of the dissection was tiresomely long, and it was deemed unwise to proceed with a gastrectomy at that time. The gall-bladder appeared normal and was not removed.

Various cases of choledochoduodenal fistula have been described, but in most the fistula has followed a calculous cholecystitis. Other causes of such fistulae include malignancy and intentional construction at operation. In the case reported by Yon and Bell (1953) retrograde filling of the biliary tree occurred after a barium meal, but these authors were careful to point out that such a filling may follow reflux through a relaxed sphincter of Oddi and is not pathognomic of a choledochoduodenal fistula. It is perhaps surprising because of its close proximity to the duodenum that the common bile duct is not more frequently involved by penetrating duodenal ulcers.

When the dissection necessary to free the stomach or duodenum in the presence of a penetrating ulcer is not too time consuming with the patient's general condition it is, of course, reasonable and indeed wise to proceed with a partial gastrectomy. Yet, in other cases, after such preliminary dissection it is necessary to keep the remainder of the operation to a minimum. Excision of the ulcer and closure of the defect alone has a definite place in the debilitated patient racked with pain, but it will not ensure freedom from the formation of a new ulcer. Theoretically, after recovery from the first operation an elective partial gastrectomy could be performed later. This arrangement has the obvious and definite disadvantage that almost invariably the patient will refuse the second operation until a new ulcer has developed and complications have presented themselves. The addition of a lesser procedure than partial gastrectomy to excision of an ulcer offers

little protection against further ulceration whether this procedure be vagotomy, gastroenterostomy, or pyloroplasty.

Because of the magnitude of the Billroth I and Polya procedures and their unsuitability for certain poor risk cases, attention is drawn to Wangenstein's descriptions (1953) of his tubular gastric resection with transverse gastropasty. This consists of a wedge resection of the greater part of the stomach and is combined with a pyloroplasty when there is any pyloric obstruction. Although not used in the above cases this tubular gastric resection with transverse gastropasty was used by me last year in 18 other poor risk patients without a fatality and, as yet, without any post-operative troubles such as the dumping syndrome. Since it is technically simpler than other types of gastrectomy it should have a lower mortality rate; and, for other reasons, it is even possible that it may have a lower morbidity rate than the modifications of the Billroth I and Polya procedures. It is therefore concluded that it is an operation worth consideration in the presence of a difficult penetrating ulcer.

#### SUMMARY

Peptic ulcers penetrating the liver, spleen, and common bile duct are described.

It is concluded that in cases of extreme difficulty the initial operative treatment of a penetrating ulcer should be limited to excision of the edges of the ulcer and closure of the resultant defect in the stomach or duodenum, a definitive gastrectomy being performed subsequently. In addition, it is suggested that in some other cases of penetrating peptic ulcer Wangenstein's tubular gastric resection with transverse gastropasty has a place as the initial procedure.

#### REFERENCES

- WANGENSTEEN, O. H. (1953), *Minnesota Med.*, vol. 36, page 924.  
— (1953), *Rev. Gastroent.*, vol. 20, page 611.  
YON, J. L. and BELL, L. G. (1953), *Arch. Surg.*, vol. 66, page 260.

## DIVERGENT STRABISMUS

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**D**URING the last ten years there has been considerable advance in our knowledge of various types of squint, due in large measure to the interest shown in this condition by surgeons in English speaking countries. However convergent squint has attracted most of the attention, probably because it is more common, more disfiguring and causes amblyopia more frequently. In consequence divergent squint has been somewhat neglected and there is no uniformity of opinion on aetiology, signs or treatment. This lack of uniformity may be linked up with the variability of the condition which is one of the most interesting and striking features seen, and it suggests that divergent squint is not a single entity but a complex of several clinically separable conditions.

In an attempt to elucidate these problems, particularly with regard to treatment, 145 cases of divergent strabismus complete with orthoptic, operative and post-operative findings have been collected and reviewed. Many other cases had to be omitted because of insufficient data, particularly in regard to the follow up, many cases being lost sight of within a few months of operation. Others had had insufficient treatment to warrant any conclusion. These 145 cases have been analyzed in an attempt to find out:—

- (1) The value of orthoptic treatment before and after operation.
- (2) The indications for operation.
- (3) The results of operation.

The first thing apparent is that the incidence of divergent squint is small when compared with convergent squint, but it appears to be higher in Australia than in England, or at least more parents bring their children along seeking advice in Australia than in England. Duke-Elder (1949) quotes Lagleyze in 1913

as giving the figures 1:4 for the relative proportion of divergent and convergent squint, but some recent English figures (Duthie, 1946) suggest 1:10 which from the author's observation is a more accurate figure.

It is more common in females than males. In this series there were 93 females and 52 males; this seems to be a universal observation.

Divergent squints do not have a common aetiology and for consideration must be split into groups. There are first those which are obviously paralytic and due to a partial or complete third cranial nerve lesion. These all require surgery if they are to be improved and will not be further considered. Divergent squints following loss of useful vision in one eye and divergence following overcorrection of a convergent squint are also excluded from this discussion, as is also divergence from space occupying lesions in the orbit.

The classification of the remaining squints numbering 145 is not easy, but from the clinical standpoint, particularly that of treatment, the following is the best (Duthie, 1946):—

- (1) Tonic.
- (2) Small angle divergence.
- (3) Divergence excess.

The characteristics of each of these types of divergent squint is as follows. The tonic is constant and has a constant angle, and is usually unilateral but can be alternating. Binocular vision is feeble and there is strong suppression. There were 27 squints that fell into this group. In all the visual acuity was 6/12 or better in each eye—usually it was 6/6, and there was usually only a small refractive error. Because of the dense suppression there was no diplopia and Worth's

lights were 3 or 2. In most cases the angle was large; being usually about -20 degrees and having -40 degrees as the upper limit and -12 degrees as the lower limit. Convergence was weak or absent, and the patient presented because of the appearance of the eyes rather than because of symptoms. All these cases received treatment, the earlier ones tending to have much orthoptic treatment while the latter were submitted to operation much sooner and often after only one visit to the orthoptic department for investigation.

The number of pre-operative orthoptic treatments in this group is extremely variable, ranging from one case that had 37 treatments to those, who are a large number, that had only one visit to the orthoptic department. This variability is accounted for by the fact that the cases were under various surgeons who had variable ideas about the use of orthoptic training and to the trend of opinion in recent years against long courses of orthoptic treatment.

Of the 27 cases one case had 37 treatments and had the angle of deviation reduced from -15 L/R 14 degrees to 0 degrees with no height difference. What the subsequent history was could not be determined, but from experience of other similar cases it is highly probable that the angle recurred. Of the remaining 26 all but 7 (i.e. 19) were submitted to operation on either one or both eyes, and the immediate results were good but there was a definite tendency for the angle to increase again after two or three years, particularly if there was a small residual angle of 5 to 10 degrees after operation. Following operation usually some orthoptic treatment was given to overcome suppression and stimulate binocular vision, but often the suppression was very deep.

The second group in the classification is the small angle divergent squint. It is characterized by a constant angle of approximately 5 degrees or less at the synoptophore, but with marked exophoria for near on the Muddox wing and poor convergence. There is usually good binocular vision except on convergence when suppression occurs. There

were four cases in this group and they presented because of symptoms of asthenopia on close work. All had good binocular vision. Their refraction and visual acuity were not noteworthy. None of the four was operated on, but all were given prolonged orthoptic treatments of 22, 13, 10 and 6 treatments with marked improvement in symptoms—but no reduction in the angle of divergence. Under conditions of fatigue and strain the condition returns but responds to a further course of orthoptic exercises and owing to the small angle and satisfactory results of orthoptic exercises operation is contra-indicated.

The remaining cases numbering 114 have been classified as divergence excess. This type of divergent squint is characterized by its variability. The angle is usually moderate, varying from 10 degrees to 30 degrees and it is voluntarily controllable. There is good convergence, and the squint is often intermittent and worse if tired. The vision and refraction are not noteworthy. The state of binocularity is variable. It is usually good but occasionally is poor. The patients present because of the appearance of the squint or because of asthenopia. Of the 114 cases in this class 30 were treated by operation with good results—and the final angle being less than 5 degrees in 24 cases. The remaining cases were treated with orthoptic exercises over varying periods. One case had 64 orthoptic treatments with reduction of the angle from -25 to -15 and relief of symptoms. However the reduction in the angle appears to be only temporary and after some months recurs. Another course of exercises will cause relief of symptoms and improvement in the angle of divergence—and in patients adverse to operation repeated short courses of orthoptics is a satisfactory method of treatment. However only operation can cure this condition.

The results of surgical interference in divergent squints are on the whole good. Poor results are usually due to an insufficient operation, and the results of overcorrection are not so much to be feared as in convergent squint. Tenotomy of the external rectis is rarely a sufficient operation. In no

case in this series where the deviation was sufficient to warrant operation, has it proved satisfactory. It should always be accompanied by resection of the medial rectus. In principle a tenotomy being an uncontrolled operation is bad and too subject to chance. A recession is a more surgical procedure and although full abduction usually returns after a tenotomy the recession operation makes this certain.

#### SUMMARY

An analysis of 145 cases of divergent strabismus is made and shows they fall in three groups. The feature of each group and its treatment is indicated.

#### REFERENCES

- DUKE-ELDER, S. (1949), "Text Book of Ophthalmology," vol. 4. London, Kimpton.  
DUTHIE, O. M. (1946), *Trans. ophthal. Soc. (U.K.)*, vol. 66, page 571.



# THE MULTIPLE GROWTH OF MOLLUSCUM SEBACEUM IN DONOR AND RECIPIENT SITES OF SKIN GRAFT

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**M**OLLUSCUM sebaceum is a common lesion in South Australia and certainly occurs elsewhere with a greater frequency than the few reports in the literature would indicate. The discrepancy is partially due to the reluctance of many clinicians and pathologists to recognize its separate identity but is mainly the result of indecision in histological differentiation from squamous cell carcinoma so that most of these lesions therefore are not allowed to prove their benign habit by being left to disappear spontaneously.

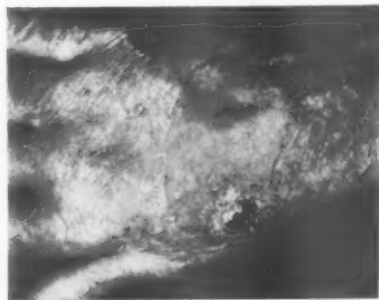


FIG. I. Shows large lesion on the radial edge of the graft, with the two other smaller lesions on the proximal and ulnar borders.

The following case of a patient with multiple skin lesions over many years is of interest because of the appearance of typical growths of molluscum sebaceum around the donor and recipient sites of skin grafts following removal of two "squamous cell carcinomas" of the back of the hand. This resulted in clinical diagnosis of malignant recurrence at the original site, and of operative implantation on to the donor area.

## CASE REPORT

The patient, a man aged 76, had been attending the Radiotherapy Department for twelve years with multiple and recurrent skin tumours of various parts of the dorsum of both hands which had been treated by surgery and irradiation. He presented in October, 1953, with two red nodules which had grown in two months on the right hand. One was in the midst of a scar of a previous lesion, healed following radiation therapy, on the dorsum of the hand; it measured 1.5 cm. in diameter. The other was a smaller lesion over the head of the fifth metacarpal bone and measured just over a centimetre in diameter. Both had bulging smooth raised edges and a central crust. These two lesions were excised, that on the back of the hand being removed (to include the scar of the previous treatment) with a large area of surrounding skin, which was approximately oval and measured 8 x 5 cm. The deficiencies were covered by split skin grafts taken from the right thigh. The pathologist reported on the two lesions as well-differentiated squamous cell carcinomas.

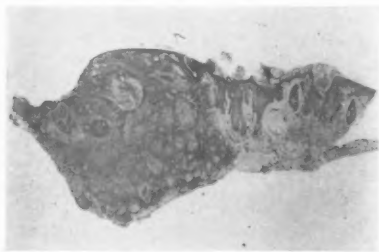


FIG. II. Biopsy from the edge of the large lesion shown in Fig. I. (x 6.5)

The grafts took well, but two months later the patient returned with further lumps, 3 on the donor site on the right thigh and 3 on the recipient site on the dorsum of the right hand. Those on the right hand (Fig. I) were all on the junction of the graft with the native skin, measuring 1.3 cm., 0.8 cm. and 0.5 cm. in diameter respectively. The lesions were raised, rounded, smooth nodules, the largest being tomato-red and having a central horny plug suggestive of molluscum sebaceum. The small graft over the head of the metacarpal bone showed

no further tumour. The lesions in the donor area were all about 1.5 cm. in diameter, one on the junction, and two within the healed zone. These were shiny, raised and rounded, but with no central crater or plug.



FIG. III. One of the tumours from the thigh. It is covered by thin squamous epithelium. One of the ducts is indicated by an arrow (see also Fig. IV). (x 7.5)



FIG. IV. On either side of the duct is a lobule of squamous epithelium undergoing keratinization. (x 100)

As molluscum sebaceum was suspected small biopsies were taken from the edge of the two larger tumours on the back of the hand, and two of those on the thigh were completely excised. The sections of the tumours on the hand showed that they were from the edge of a crateriform tumour (Fig. II) the outer surface being covered by stretched but otherwise normal epidermis, the inner lining consisting of a mass of folded burrowing well-differentiated squamous epithelium from which thick keratinous plugs protruded on to the surface. The lesions from the thigh were unusual. They consisted of lobulated-looking club-shaped downgrowths of perfectly differentiated squamous epithelium and were covered by a layer of thin epidermis (Fig. III). In one the dermal downgrowths were continuous with the surface epidermis at only two points, and these connections consisted of small duct-like pores which led to the exterior from the most superficial tumour lobules which were expanded by central keratin formation (Fig. IV). The appearance of these pores is not unlike that of a sebaceous duct. The other thigh tumour had three larger eccentric openings leading from almost cystic expansions of squamous epithelium filled with keratin flakes (Fig. V), but

one half of the surface was uninterruptedly covered by flattened epidermis. It can also be seen from Fig. V that keratinization is most advanced where communication with the surface is widest.

In none of the above tumours was there any nuclear pleomorphism, and mitotic activity was no more than is found in actively growing epithelium. It can be seen from Figs. III and V, that despite the amount of proliferating squamous epithelium, the lesion is a protuberant one, expanding the epidermis more than infiltrating the dermis. About four-fifths of its bulk are above the probable previous level of the epidermis.



FIG. V. Another lesion from the thigh showing keratinization and surface communication, both eccentric. (x 6)

On further examination of the two original tumours excised from the hand it was decided that these too were examples of molluscum sebaceum, being crateriform with keratotic plugs, very well-differentiated and superficial (Fig. VI). Re-examination of biopsies taken over the previous twelve years showed that most of these were typical examples of molluscum sebaceum, the remainder being uncertain, due to unfavourable biopsy material, but very consistent with this diagnosis.



FIG. VI. The original larger lesion from the hand, showing hyperplastic epithelium and some surface keratinization. (x 5)

### Progress

The remaining lesion on the thigh had disappeared in two months and there were no recurrences. The two smaller lesions on the hand had disappeared in

three months, but the large one persisted, appearing to heal on the medial side while extending on the lateral side.

In August, 1954, a further red rounded nodule 1.4 x 1.3 cm. with a central horny plug, appeared in the skin of the right wrist just outside and proximal to the graft, and was considered to be a molluscum sebaceum. This lesion and the unhealed one on the rim of the graft were given small doses of X-rays with slow resolution. The latter (Fig. VII) is represented, 12 months from its first appearance, by a nodular area of thickened skin, approximately 1 cm. in diameter. The recent nodule, after 5 months, is slightly smaller but still has a central horny plug. No further nodules have appeared.



FIG. VII. Remnants of the large lesion on the radial edge of the graft, but the other two lesions have resolved without treatment.

### DISCUSSION

As the features of molluscum sebaceum have recently been described by Beare (1953) and by Fouracres and Whittick (1953) it will not be necessary to enter upon a general description of the disease here. Fouracres and Whittick have pointed out the probable identity of molluscum sebaceum and multiple self-healing squamous cell carcinoma of the skin. All the evidence clearly supports their view and under different circumstances it is easy to see how the present case could have been classed as a self-healing "malignancy."

The multiple character of the present lesions, and particularly their appearance in a distant donor graft site, may be interpreted as evidence of an infective process. On the other hand the circumstances may indicate a traumatic basis, sequestration of epithelium being followed by proliferation. In this regard it should be noted that Beare was unable to produce tumour formation by intradermal injection of crude filtrates into his own forearm.

From a consideration of the lesions on the thigh it is possible to sketch the evolution of molluscum sebaceum, the early stages of which have been relatively neglected in the literature. One of the lesions on the thigh communicated with the exterior by microscopic pores (two being found) which led from an expansion of epithelium filled with keratin. By the increasing production of keratin in the submerged epithelium it is probable that the pores become widened and the keratotic plug is slowly extruded, not unlike the unfolding of a sea anemone. If the pore-like communications are eccentrically placed the tumour may appear to heal on one side and extend on the other side as did the largest tumour on the back of this patient's hand (see also Fig. V).

A further point which arises from this case is the apparent risk of inflicting further lesions by unnecessary surgical action, a possibility suggested recently by the experience of Beare (1955). It is important, therefore, to become familiar with the appearance of molluscum sebaceum so that radical measures may be put aside with confidence. Curetting the lesion at the everted keratotic stage gives very satisfactory material for microscopy and at the same time the ease with which the mass shells out usually gives some indication of its non-infiltrative nature.

### SUMMARY

Following the removal of a molluscum sebaceum from the back of the hand of a male aged 76, further tumours appeared, in two months, on the donor and recipient sites of skin graft. Two were completely excised and one is regressing; the other lesions disappeared spontaneously.

### ACKNOWLEDGEMENTS

We would like to thank the Royal Adelaide Hospital Board, Dr. B. S. Hanson and Dr. J. R. Magarey for permission to publish this case. For advice and help with the photography we are indebted to Professor J. S. Robertson and Mr. W. Nolan, the latter providing the two clinical photographs.

### REFERENCES

- BEARE, J. M. (1953), *Brit. J. Surg.*, vol. 41, page 167.
- (1955), *Lancet*, vol. 1, page 182.
- FOURACRES, F. A. and WHITTICK, J. W. (1953), *Brit. J. Cancer*, vol. 7, page 58.

## Books Reviewed

### THE DIAGNOSIS OF THE ACUTE ABDOMEN IN RHYME.

By ZETA. London: H. K. Lewis & Co., 1955. Third Edition. 7½" x 5", vii plus 96 pp., illustrated. Price: 7s. 6d.

The appearance of a third edition of this volume of surgical diagnosis in rhyme speaks well for its popularity. It was first published in 1947 and the second edition appeared in 1949. Written by a distinguished English surgeon the book is full of sound advice and enriched by the wide experience of its author. The drawings by Peter Collingwood are full of humour.

### WAR SURGERY AND MEDICINE.

By T. DUNCAN M. STOUT, F.R.C.S., F.R.A.C.S. Wellington, New Zealand: R. E. Owen, Government Printer, 1954. 9½" x 6", xiv plus 779 pp., 46 illustrations, 3 maps.

This volume is part of the official history of New Zealand in the Second World War, 1939-45, and it is the first of three volumes covering the medical history of New Zealand in this war. It is a clinical volume reporting the most important aspects of the clinical work and experience of the New Zealand Medical Corps over this period. The author is well suited to deal with his subject as he served during World War I (1914-1918) as a specialist surgeon, while throughout World War II he was the Consultant Surgeon to 2nd N.Z.E.F. and so was able to assess at first hand the problems of war surgery as they arose.

The book is divided into two main parts—the Surgical Section and the Medical Section, and the record of experiences covers the campaigns in the Middle East, in Italy, and in the Pacific, and the lessons drawn are from a survey of casualties which include 1,464 men who died of wounds (excluding those killed in action), 16,475 wounded and over 100,000 sick.

The Surgical Section opens with a survey of wound treatment and contrasts the methods of World War I and the Spanish Civil War, with methods as they evolved during World War II. The steady improvement in results during successive campaigns, as earlier and more efficient surgery made for a considerable decrease in wound sepsis, compares vividly the closed plaster techniques of the earlier Libyan campaigns with the possibilities of delayed primary suture of wounds during the later phases of the war in Italy.

Next follows a discussion on forward surgery. The development of the Field Surgical Units and the Specialist Units to cope with the problems of neuro surgery, of maxillo facial surgery and of ophthalmology are outlined. The problems of evacuation which bear on the "Level" at which forward surgery is best performed, the selection of cases in their order of priority for the forward surgeons and the clinical features of these cases are well discussed.

The following twelve chapters are then each devoted to a special type of wound. Abdominal injuries, chest injuries, head injuries, fractures and amputations, etc., are discussed in turn and statistics and results are commented on. Further chapters

cover such problems as foot disabilities, hernia, varicose veins and haemorrhoids and their affect on the boarding and grading of the soldier.

The Medical Section is divided into three parts—(1) Infective Diseases, (2) Systemic and Constitutional Diseases, (3) General. The section on Infective Diseases has seventeen chapters, each of which is devoted to a specific problem. Dysentery, infective hepatitis, malaria, venereal disease, etc., with their control and their bearing on the health of the force are each discussed in detail and again statistics are provided. In the control of pulmonary tuberculosis, the importance of routine chest radiography on enlistment and initial boarding is shown.

In the section dealing with Constitutional Diseases there are interesting chapters on dyspepsia, neurosis, essential hypertension and skin diseases together with useful comments by the Medical Officer in Charge of the War Pensions Department on the disability factors of these diseases.

Colonel Stout is to be congratulated on the production of this work. It is well produced, and it supplies an accurate and useful record of the medical and surgical problems as they arose in the various campaigns of World War II. It will thus be read with interest by those who have been called to deal with the problems of war surgery and medicine and it will serve as a useful reference book for future medical officers.

### DEMONSTRATIONS OF PHYSICAL SIGNS IN CLINICAL SURGERY.

By HAMILTON BAILEY, F.R.C.S., F.A.C.S., F.I.C.S., F.R.S.E., assisted by ALLAN CLAIN, M.B.(Cope Town), F.R.C.S.(Eng.). Twelfth edition. Bristol, Eng.: John Wright and Sons Ltd., 1954. 8½" x 5½", xii plus 456 pp., 681 illustrations. Price: 38s. 6d.

This book is aptly named "Demonstrations of Physical Signs in Clinical Surgery," as the text is so well illustrated by clinical photographs and diagrams that the student can have little difficulty in understanding the methods of examination which are described.

Mr. Hamilton Bailey has been assisted in the preparation of this twelfth edition by Mr. Allan Clain, and the authors have effected still further improvement in the range and quality of the illustrations, although in previous editions this feature left little to be desired. There are a few minor inaccuracies in the text, but the book is generally of such a high standard that it remains an essential one for students and one which the clinical teacher may consult with advantage.

## Book Received

### L'IPOTERMIA GENERALE CONTROLLATA IN CHIRURGIA.

By PIERO GOFFRINI and EUGENIO BEZZI. Itchy: Omnia Medica, 1954. 9½" x 6½", vii plus 194 pp., 51 illustrations. Price: L.2000.

### THE ROLE OF THE PITUITARY IN CANCER.

By HENRY K. WACHTEL, M.D. New York, U.S.A.: The Williams-Frederick Press, 1954. 8½" x 5½", 31 pp. Price: \$2.00.

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